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Rare Variation in Partial Anomalous Venous Drainage in 2 Cases: Diagnosis, Assessment Methods, and Surgical Approach

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ABSTRACT

Where pulmonary veins drain and their relationship with an atrial septal defect (ASD) are important. A sinus venosus (high venosum) type of defect is the most common pathology accompanying partial anomalous pulmonary venous connection. Typically, the right superior pulmonary vein and occasionally the middle pulmonary vein drain into the junction of the superior vena cava (SVC) and the right atrium (RA), and a sinus venosus type of ASD usually accompanies these anomalies. In this report, we assess a very rare pathology in which 3 right pulmonary veins (superior, middle, and inferior) drain into the SVC-RA junction with respect to diagnostic methods and in the light of 2 cases involving patients in 2 different age groups.

INTRODUCTION

A partial venous connection anomaly is a failure of one or more pulmonary veins to connect to the left atrium during fetal development. Such anomalies can vary according to the site of drainage of the pulmonary veins: drainage of the right pulmonary vein into the junction of the superior vena cava (SVC) with the right atrium (RA); drainage of the right pulmonary vein directly into the RA and the inferior vena cava (scimitar syndrome); drainage of the left pulmonary veins into the left innominate vein; and, very rarely, bilateral partial anomalous pulmonary venous connection (PAPVC) with intact atrial septum, drainage into the junction of the right superior pulmonary vein with the SVC and RA, and junction between the left superior pulmonary vein and the left innominate vein. Although this pathology has rarely been defined in clinical series, the incidence is 0.6% to 0.7% in autopsy studies. Despite the excellent early surgical outcomes in various age groups, SVC obstruction and rhythm disorders are the most important postoperative problems [Kirklin 1993]. Another issue is the role of noninvasive diagnostic methods in the assessment of patients with this pathology [Kafka 2009].

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CASE REPORTS

Case 1

An 8-year-old girl was admitted to our pediatric cardiology unit for a cardiac murmur. An electrocardiogram revealed an incomplete right bundle branch block and signs of RA and right ventricular volume load. A transthoracic echocardiography (TTE) examination indicated dilation of the RA, the right ventricle, and the main pulmonary artery, in addition to an atrial septal defect (ASD) (sinus venosus type) and a PAPVC with the right pulmonary veins emptying into the right side of the SVC. The left pulmonary veins drained normally. The echocardiography evaluation also revealed that the left-sided SVC drained into a markedly enlarged coronary sinus. A 64-slice multidetector computed tomography (MDCT) scan showed PAPVCs: right superior, middle, and inferior pulmonary veins connecting to the right side of the SVC (Figures 1A and 1B). Heart catheterization and angiography evaluations were not performed. Surgical correction was planned for the large shunt and volume load.

Case 2

A 65-year-old female patient with chest pain and dyspnea presented to the emergency department. The physical examination was unremarkable except for a 4/6 systolic murmur and grade 2+ pretibial edema. A partial-type anomalous venous drainage (superior, middle, and inferior pulmonary veins opening onto the SVC and RA junction with separate orifices) and a sinus venosus type of ASD were detected by echocardiography and the consequent MDCT investigation (Figure 1C). Coronary angiography and cardiac catheterization evaluations detected 90% stenosis at the first diagonal branch and a left-to-right shunt (Qp/Qs) ratio of 3.2.

During the operation, we noticed in the inspection that all 3 right pulmonary veins opened into the SVC-RA junction and visualized a third anatomic chamber at the site where the

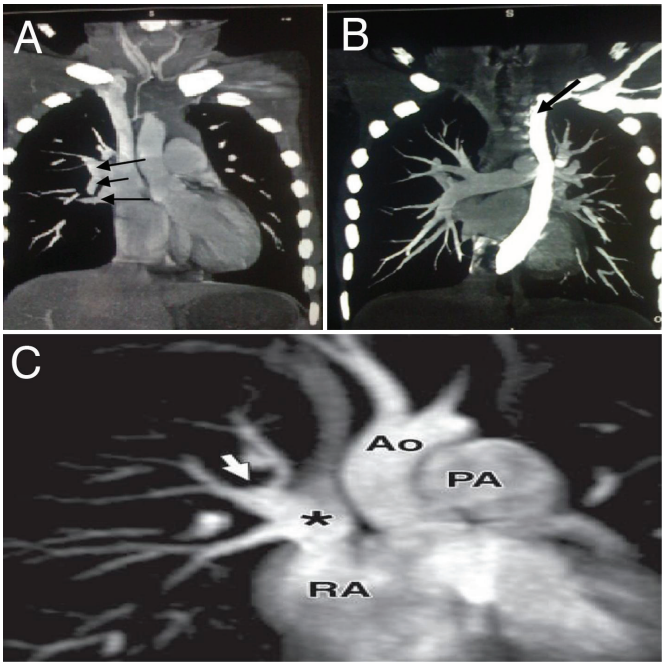

Figure 1. Preoperative cardiac multidetector computed tomography. A, Three pulmonary veins opening into the right atrium (RA) near the superior vena cava (SVC) (black arrows). B, Left persistent SVC opening into the coronary sinus (black arrow); notice the absence of the innominate vein. C, Third chamber formed by 3 pulmonary veins (*) opening into the SVC-RA junction. Ao indicates aorta; PA, pulmonary artery; RA, right atrium.

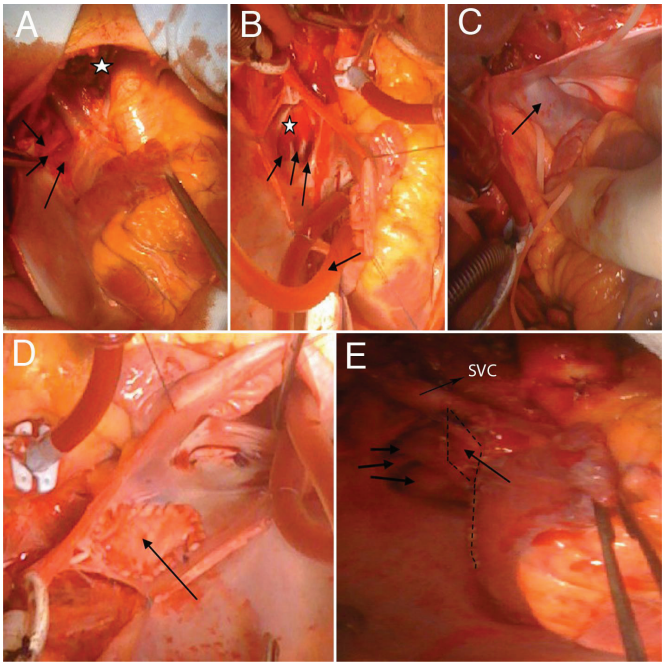

Figure 2. A, Pulmonary veins draining into the third chamber and the superior vena cava (SVC) (black arrows). Note the absence of the innominate vein (star). B, Entrance orifices of 3 pulmonary veins, the third chamber (star), and the vent in the coronary sinus. C, Left persistent SVC. D, Closure of the sinus venosum with pericardial patch so that the pulmonary veins remain in the left atrium (arrow). E, Widening of the entrance orifice of the SVC to the right atrium with pericardial patch (arrow) and pulmonary veins.

pulmonary veins opened. A left persistent SVC (LPSVC) was present, and the innominate vein was absent (Figures 2A and 2C).

Operative Technique

The operative technique was as follows: The thorax was opened by median sternotomy. A piece from the anterior pericardium was prepared to use as a graft and was treated with 0.6% glutaraldehyde. After heparinization, aortic and bicaval venous cannulation was performed, followed by SVC, inferior vena cava, and LPSVC taping. The left SVC pressures measured for the 2 cases were 10 mm Hg and 12 mm Hg, respectively. When the RA was opened, the placing of a sump sucker vent (Medtronic, Minneapolis, MN, USA) was concluded, and extracorporeal circulation (ECC) was initiated. After clamping of the aorta, cardiac arrest was provided by blood cardioplegia. Patients were cooled to 26°C. In ECC via an RA approach, 3 pulmonary veins opening near the junction of the SVC and the RA drained into the left atrium via the sinus venosus type of ASD. The defect was closed with a pericardial patch (Figure 2D). During the RA closure, widening was achieved with the pericardial patch to prevent obstruction at the SVC-RA junction (Figure 2E). Single-vessel coronary artery bypass surgery (aorta to first diagonal branch) was performed in the 65-year-old patient. Both patients emerged from ECC in sinus rhythm and with no complications. The postoperative periods were unremarkable, and the patients were discharged on the eighth postoperative day.

DISCUSSION

Use of TTE to assess the variations in the pulmonary vein anatomy of patients with abnormal pulmonary venous drainage is a challenge. The width, the trace, and the drainage site are important for the pulmonary veins. These veins may sometimes be at a distance from the heart, which makes it impossible to define them accurately. Although the rate of an accurate diagnosis with TTE performed in a fetus with a congenital heart defect is 85% to 90%, the presence of an abnormal aortic arch and abnormal pulmonary veins may be overlooked [Forbus 2004]. An adequate acoustic window for investigation may not be found in some cases [Wong 1995]. Therefore, the pulmonary venous anatomy should be assessed so as to not encounter a surprise, or prior to the transcatheter closure procedure [Cragun 2005].

PAPVC occurs in 10% to 15% of patients with secundum ASDs and in 85% of patients with sinus venosus ASDs. In our cases, there was a very rare PAPVC anomaly with an accompanying sinus venosus type of defect with 3 right pulmonary veins (superior, middle, and inferior) draining into the SVC-RA junction. In this complicated situation, it is difficult to make an accurate diagnosis with routine TTE. Therefore, other imaging techniques are necessary for a comprehensive diagnosis of PAPVC. Although cardiac catheterization has been the method of choice in the past, transesophageal

echocardiography (TEE), CT, and magnetic resonance imaging (MRI) have replaced it, because TTE is invasive and causes ionizing radiation. In cases in which left-sided pulmonary veins drain into the SVC from a higher site, the success rate of TEE is lower. On the other hand, one should keep in mind that CT causes ionizing radiation and that MRI requires sedation in children [Modan 2000; Kleinerman 2006]. Thus, MRI seems to be the most suitable method for visualizing the pulmonary vein anatomy and for measuring shunts from left to right [Grosse-Wortmann 2007]. Although pulmonary vein anomalies have traditionally been evaluated with TTE and angiography, they can be accurately diagnosed with both MRI and MDCT [Dillman 2009]. We successfully used 64-slice CT in our 2 patients for preoperative diagnosis. The rare pathology of complete right pulmonary veins opening into the SVC-RA junction to form a third chamber was assessed in detail with this method. Although rare, their placement just opposite the sinus venosus type of ASD has allowed an ideal surgical intervention. Otherwise, the cavoatrial anastomosis defined by Warden in 1984 is used in the pathology of high pulmonary vein entry into the SVC, which renders the surgery complicated [Warden 1984]. The most important problem here is the cavoatrial stenosis due to SVC tension. The recommended ideal surgical technique is closure of the ASD and directing the pulmonary veins to the left atrium with a pericardial patch. The important point is to enlarge the narrowed SVC-RA junction again with pericardium. This strategy will avoid obstruction and rhythm problems. We have had successful outcomes with this technique, which we used in these 2 patients. Another important consideration is applying specific cannulation techniques when correcting these sporadic cardiac pathologies. In particular, if there is a left SVC and no innominate vein connection and the venous pressure occlusion test indicates a pressure >18 mm Hg, such a system should be cannulated directly. If there is a medium-sized left SVC thought to be carrying significant flow, it should be returned to where it opened to the heart, and with a little incision in the sac suture (without occluding the aperture), the free drainage to the pericardial cavity during ECC should be evaluated. Alternatively, as in our cases involving the presence of a LPSVC, a sump might be placed in the coronary sinus. Thereby, surgical comfort and success are gained.

The patients were assessed at postoperative month 8 by MRI and were found to have no problems. In conclusion, characterization of rare variations in PAPVC anomalies, which is important in deciding the surgical strategy, is easily accomplished with additional imaging techniques, and such anomalies, once identified, are treated successfully with the optimal surgical intervention.

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