Double drainage of total anomalous pulmonary venous connection: A rare variant of mixed total anomalous pulmonary venous connection

Mani Ram Krishna[1], Ganesh Kumar Gnanappa[1], Jonathan Mervis[1], Jonathan Forsey[1], Julian Ganesh Ayer[1], Yishay Orr[2]
[1]Department of Cardiology, The Children's Hospital at Westmead, Westmead, Australia, [2]Department of Cardiothoracic Surgery, The Children's Hospital at Westmead, Westmead, Australia

ABSTRACT

Total Anomalous Pulmonary Venous Connection (TAPVC) is frequently classified based on the system proposed by Craig, Darling and Rothney. Occasionally variants are reported which do not fit into these classic forms. One such variant is the double drainage of TAPVC where the confluence has connections at two different levels. We report two infants with a double drainage pattern of TAPVC.

Keywords: Cyanosis, total anomalous pulmonary venous connection, unobstructed drainage

Total anomalous pulmonary venous connection (TAPVC) was classified by Craig *et al.*[1] into supracardiac, cardiac, infracardiac, and mixed types based on only 17 autopsies. Due to the small sample size, it is inevitable that variants of the venous connections exist which do not fit into the classification. A rare variant is TAPVC with double drainage[2] where a common chamber has connections at multiple levels to the heart. We describe two patients with a variant of the double drainage pattern.

CLINICAL DETAILS

Case 1

A 3-day-old baby was referred for the evaluation of persistent desaturation. An echocardiogram suggested TAPVC with double drainage where the pulmonary veins formed a common chamber behind the heart which drained into the innominate vein through an ascending vertical vein [Figure 1] as well as directly to the coronary sinus. A cardiac computerized tomogram (CT) angiogram [Figure 2] showed that the right-sided pulmonary veins drained into a transverse confluence behind the left atrium (LA) whereas the left-sided pulmonary veins drained into a separate vertical confluence lateral to the LA appendage. The right-sided pulmonary venous confluence drained directly to the coronary sinus. In addition, there was a small communicating channel between the right-sided confluence and the left-sided confluence which then drained through an ascending vein to the innominate vein. These findings were confirmed intraoperatively.

Case 2

A 4-day-old newborn with an uneventful perinatal history was referred for persistent desaturation. An echocardiogram identified anomalous pulmonary venous return to a common chamber posterior to the LA, a large left-sided ascending vertical vein to the innominate vein and an additional ascending vertical vein draining directly to the right superior vena cava. Both the veins were unobstructed. CT confirmed the pulmonary venous drainage pattern [Figure 3]. The anatomy at surgery was as described in the preoperative imaging.

Access this article online

Quick Response Code:

Website: www.annalspc.com

DOI: 10.4103/apc.APC_45_19

How to cite this article: Krishna MR, Gnanappa GK, Mervis J, Forsey J, Ayer JG, Orr Y. Double drainage of total anomalous pulmonary venous connection: A rare variant of mixed total anomalous pulmonary venous connection. Ann Pediatr Card 2020;13:100-1.

Address for correspondence: Dr. Mani Ram Krishna, The Heart Centre for Children, The Children's Hospital at Westmead, Locked Bag 4001, Westmead NSW 2145, Australia. E-mail: mann_comp@hotmail.com

Received: 01-04-2019 **Accepted:** 03-04-2019 **Published:** 19-07-2019

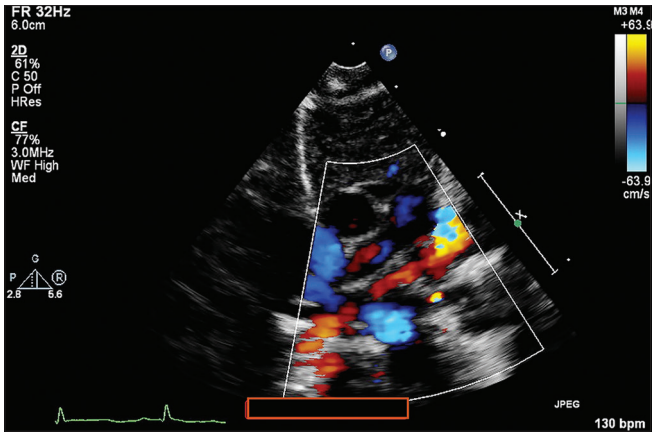

Figure 1: Two-dimensional transthoracic echocardiogram with color Doppler showing drainage of pulmonary veins into a confluence with communications to an ascending vein as well as to the coronary sinus

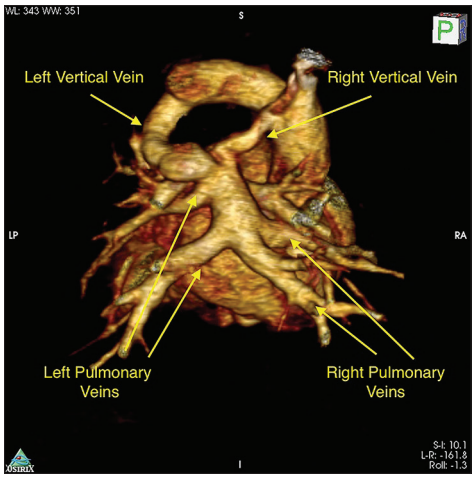

Figure 3: Three-dimensional volume-rendered images from the cardiac computerized tomogram viewed from the posterior side. The pulmonary veins drain into a confluence which then communicates to both the innominate vein and the superior vena cava through two separate ascending vertical veins

DISCUSSION

Double drainage of TAPVC is a rare variant of mixed TAPVC. TAPVC arises from failure of the common pulmonary vein to incorporate into the LA (around 30 days' gestation). This results in persisting connections between the primitive pulmonary venous plexus and the cardinal venous systems. It is purported that double drainage TAPVC results from an earlier embryological insult, resulting in persistence of communications at multiple levels.[3] This is similar to the classical mixed pattern, and the specific embryological triggers for this variant have not been identified.

Our first case is distinctive due to the presence of separate common chambers for right and left pulmonary veins, a communication between these chambers and the coronary sinus and innominate vein. To the best of our knowledge, this unusual variant has not been previously

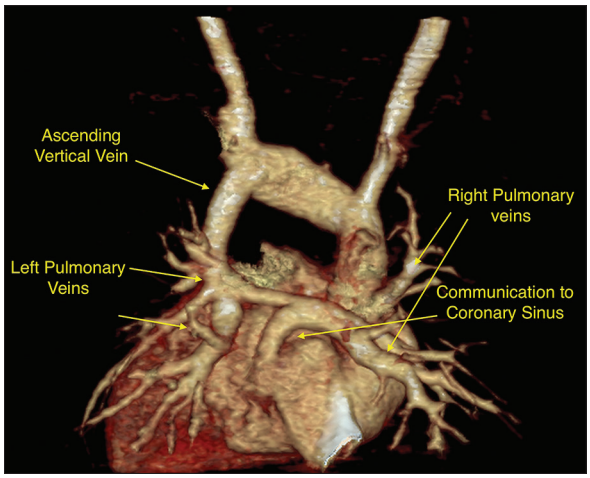

Figure 2: Three-dimensional volume-rendered images from the cardiac computerized tomogram viewed from the posterior side. This shows the pulmonary veins of each side draining into separate confluences with a communication between the two. The pulmonary venous blood enters the systemic circulation through both the coronary sinus and an ascending vertical vein to the innominate vein

reported. Echocardiography is adequate to delineate the anatomy for the common variants of TAPVC. CT angiography (with three-dimensional data) provides more detailed assessment of complex pulmonary venous anatomy allowing better preoperative planning.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Craig JM, Darling RC, Rothney WB. Total pulmonary venous drainage into the right side of the heart; report of 17 autopsied cases not associated with other major cardiovascular anomalies. Lab Invest 1957;6:44-64.
2. Arciprete P, McKay R, Watson GH, Hamilton DI, Wilkinson JL, Arnold RM. Double connections in total anomalous pulmonary venous connection. J Thorac Cardiovasc Surg 1986;92:146-52.
3. Tanabe S, Nakasato M, Suzuki H, Ishikawa A, Fukasawa M, Hayasaka K. A new form of total anomalous pulmonary venous connection with double drainage. Pediatr Int 2000;42:369-71.