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

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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.

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**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. 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 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.

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**Conflicts of interest**

There are no conflicts of interest.

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