Images in Pediatric and Congenital Heart Disease

Rare Cases of Mixed Type Total Anomalous Pulmonary Venous Connection with Chambers Encircling Left Atrium

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Mixed type total anomalous pulmonary venous connection has the morphologic heterogeneity of the venous connection. The anatomical subtypes of mixed type total anomalous pulmonary venous connection were classified by Chowdhury and colleagues; bilateral and symmetrical connections, bilateral but asymmetrical connections, bizarre anatomic variants.[1, 2)] We described two infants of mixed type total anomalous pulmonary venous connection with chambers encircling left atrium. We diagnosed by 80-slice helical computed tomography (Aquilion PRIME; Canon).

Case 1

A six-day-old, term, male infant presented with cyanosis. We diagnosed total anomalous pulmonary venous connection and atrial septal defect. Echocardiography showed pulmonary veins forming a common chamber that connected to the innominate vein; however, the

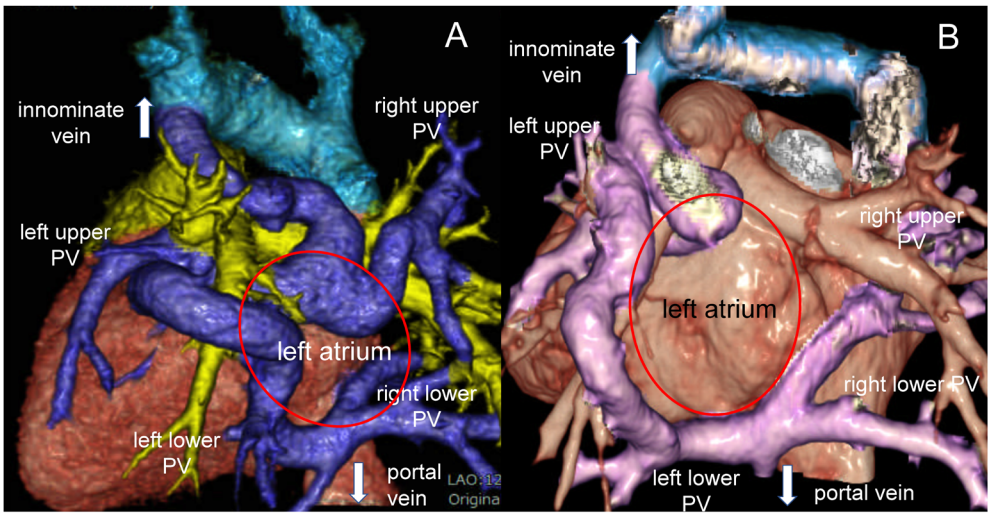

Fig. 1 Cardiac computed tomography shows pulmonary veins form chambers encircling left atrium (red circle) which drain into innominate vein and portal vein in Case1 (A), in Case 2 (B)

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drainage path of the right pulmonary veins could not be identified. Cardiac computed tomography (CT) confirmed that the right lower and left pulmonary veins formed the chamber ascending into the left lung, and partially draining into the portal vein. It joined the right upper pulmonary vein, and connected to the innominate vein (Fig. 1A). The chamber was directly anastomosed to the left atrium on 8 days old. All pulmonary veins were patent 1 year postoperatively.

Case 2

A one-day-old, term, male infant presented with cyanosis. We diagnosed total anomalous pulmonary venous connection and atrial septal defect. Echocardiography revealed drainage of the vertical veins to both the innominate and portal veins, however connection between the pulmonary and vertical veins was not fully identified. Cardiac CT confirmed that the right and left lower pulmonary veins formed a common chamber, that ascended behind the left pulmonary artery, and partially descended to the portal vein. It joined the left upper pulmonary vein, and connected to the innominate vein (Fig. 1B). Primary sutureless repair of the chamber of the right and left lower pulmonary veins was performed. The left upper pulmonary vein was directly anastomosed to the left atrium on 3 days old. Three of the pulmonary veins were patent 3 years postoperatively. However, the direct anastomotic site was obstructed. The left upper pulmonary vein flow returned to the left atrium via not the direct anastomotic site but the left lower pulmonary vein.

Comments

The diagnostic difficulties with echocardiography are associated with pulmonary vein variations of mixed type total anomalous pulmonary venous connection. Cardiac CT provides accurate anatomic delineation for preoperative planning. Encircling chambers are classified as bizarre anatomic variants of mixed total anomalous pulmonary venous connection by Chowdhury,[1, 2] and are similar to previously reported long tortuous chambers.[3] However, this pattern of chambers has never been reported to the best of our knowledge. The postoperative courses were good in two cases. However, one of the two anastomotic sites was obstructed in case 2. It is important how we anastomose for this chamber to prevent pulmonary vein stenosis, and further research is needed.

Conflicts of Interest

The authors have no conflicts of interest to declare.

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