A Case of Total Left Anomalous Pulmonary Venous Connection with Intact Atrial Septum Diagnosed by Two-Dimensional and Doppler Echocardiography

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SUMMARY

Unilateral total anomalous pulmonary venous connection from a unilateral lung is extremely rare. A 6-year-old patient with anomalous pulmonary venous connection from the entire left lung to the left innominate vein, with an intact atrial septum, diagnosed by two-dimensional and Doppler echocardiography is reported. The combination of two-dimensional and Doppler echocardiography is very useful not only for anatomical diagnosis, but also for evaluation of its hemodynamics, despite the wide anatomical variability of the pulmonary venous connection. This is the first report of a case of left total anomalous pulmonary venous connection diagnosed noninvasively.

Additional Indexing Words:
Anomalous pulmonary venous return  Unilateral total anomalous pulmonary venous connection  Echocardiographic diagnosis
Anatomical variability  Hemodynamics  Atrial septal defect

A nomalous pulmonary venous return represents a relatively rare congenital anomaly with wide anatomical variability.\textsuperscript{1)} It is occasionally associated with other cardiac anomalies, particularly atrial septal defect,\textsuperscript{1)} and may therefore present a difficult problem in the precise diagnosis and evaluation of its hemodynamics. Cardiac catheterization and angiography have been performed to quantitate the shunt size and to accurately demonstrate the site of the pulmonary venous return and the atrial septal defect. We recently diagnosed an anomalous pulmonary venous connection from the entire left lung to the left innominate vein with an intact atrial septum, by two-dimensional and Doppler echocardiography, and herewith describe the findings.
A 6-year-old girl was admitted to our hospital for evaluation of a cardiac murmur detected at routine medical examination in elementary school. She had been asymptomatic. Physical examination revealed normal growth (height: 119.5 cm, weight: 17.6 kg). The patient's blood pressure was 100/58 mmHg, pulse rate regular at 78/min and respiratory rate 16/min. On auscultation, a grade 2/6 soft systolic ejection murmur with a faint diastolic murmur was audible at the upper left sternal border. The first sound was normal, and the second sound was widely split throughout the respiratory cycle. The chest roentgenogram demonstrated slightly increased pulmonary vascular markings and a protruding left second arch without cardiac enlargement. The electrocardiogram demonstrated an incomplete right bundle branch block and right ventricular overload with right axis deviation.

The two-dimensional (2-D) echocardiogram demonstrated right atrial and ventricular dilatation with a paradoxical septal motion. An atrial septal defect (ASD) was not detected. In the suprasternal approach, the dilatated innominate vein (I.V.), vertical vein (V.V.) and superior vena cava (SVC) were clearly detected (Fig. 1). The abnormal blood flow from the left pulmonary vein (PV) to the I.V. and SVC through the V.V. was also clearly demonstrated by Doppler and Color-Doppler echocardiography. The abnormal right pulmonary venous connection was not demonstrated by either 2-D or Color-Doppler echocardiography. The ratio of pulmonary to systemic blood flow noninvasively evaluated by Doppler echocardiography was 2.2.

Cardiac catheterization confirmed the left-to-right shunt with a Qp/Qs of 2.17. The oxygen data placed the shunt at the level of the left I.V. Right and left heart pressures were normal. Selective left pulmonary arterial (PA) angiography confirmed an anomalous PV connection from the entire left lung to the left I.V. through the V.V. (Fig. 2). The right PA angiogram demonstrated a normal venous return to the left atrium from the right lung. No flow of contrast across the ASD could be detected by selective right PA angiography. The patient underwent surgical correction of the anomalous pulmonary venous connection, and her postoperative course was uneventful.

Unilateral total anomalous pulmonary venous connection (UTAPVC) from either the left or right lung is extremely rare, and there are only about 8 cases of UTAPVC from the left lung, with intact atrial septum, in the
literature. In the UTAPVC with intact atrial septum, any disorder or respiratory dysfunction of the normally draining lung may be fatal because only one lung is draining oxygenated blood into the left atrium and the anomalously draining blood flow approximates 66% of the pulmonary blood flow.\(^{11,5}\) Therefore, in the UTAPVC with intact atrial septum, an accurate diagnosis is necessary and surgical correction recommended.
Fig. 2. Venous phase of selective left pulmonary arterial angiogram demonstrates that the veins from the left lung drain into a vertical vein (V.V.) which connects to the innominate vein (I.V.).

Cardiac catheterization and selective PA angiography have been necessary for the accurate demonstration of the venous return, evaluation of the shunt size and exclusion of the presence of an atrial septal defect. The recent development of echocardiographic techniques to detect cardiac anomalies provides a convenient noninvasive method not only for the anatomical diagnosis, but also for the evaluation of the hemodynamics of cardiac anomalies. As demonstrated in our case, two-dimensional and Doppler echocardiography can reveal the anatomy of an anomalous venous connection to the left innominate vein and evaluate the shunt size. The combination of two-dimensional and Doppler echocardiographic techniques aids in the accurate anatomical diagnosis, despite the wide anatomical variability of the pulmonary venous connection in UTAPVC.

References

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ECHOCARDIOGRAPHIC DIAGNOSIS OF UNILATERAL TAPVC
