Right Pulmonary Vein Atresia in a Mildly Symptomatic Boy: Comprehensive Analysis of Flow Dynamics Using Non-contrast-enhanced 4D Flow MR Imaging

Marie-Luise Kromrey, Hiroaki Kise, Junji Hirose, Masashi Yoshizawa, Takako Toda, Yosuke Kono, Yuto Sunaga, Hisashi Johno, Satoshi Funayama, Hiroshi Onishi, Kevin Johnson, Oliver Wieben, and Utaroh Motosugi

We present multimodal imaging in the rare case of isolated unilateral pulmonary vein atresia in a 6 year-old boy, including analysis of hemodynamics by magnetic resonance acquisition technique of time-resolved three-dimensional phase contrast imaging (4D flow magnetic resonance imaging). This novel imaging method enables the quantification and especially comprehensive visualization of blood flow patterns, even in complex congenital anomalies which abducted detailed assessment so far, and therefore constitutes a promising alternative to conventional vascular imaging techniques.

Keywords: unilateral pulmonary vein atresia, magnetic resonance imaging, 4D flow

Unilateral atresia of the pulmonary vein (PV) represents a scarce congenital anomaly, the isolated appearance without associated cardiac disease is even more unusual with only few cases described in the literature. Characteristic is a high morbidity and mortality caused by complications during childhood, such as pulmonary hypertension, recurrent pulmonary infections and hemoptysis due to the rupture of bronchial varices. Standardized diagnostic procedure includes cardiac catheterization, computed tomography (CT) angiography, cardiac magnetic resonance imaging (MRI) and bronchoscopy. Management options range from follow-up, coil embolization of systemic arterial collaterals and pneumectomy, the primary aim being to prevent the development of pulmonary hypertension. In cases of diagnosis in infancy, surgical intervention via pulmonary vein reconstruction has also been described.

A 6-year-old male presented with dysphagia and weight loss since 2 months. An externally performed contrast-enhanced CT showed a narrowing of the right PV and the child was referred to our institution for further investigation of cardiovascular morphology and circulatory dynamics.

Cardiac CT (Fig. 1) confirmed right PV atresia and additionally showed multiple collateral blood flow paths from the descending aorta to the right lung. Stroke volumes in the main pulmonary artery (PA) were 2013 mL/min and in the left PA 2346 mL/min. The right lung appeared volume reduced and congestive. The child had not experienced pulmonary or cardiac symptoms so far. Upper gastrointestinal endoscopy, as well as bronchoscopy yielded no pathological findings. Lung perfusion scintigraphy revealed a pulmonary blood flow ratio left:right of 91.8:1.9 and almost no detection of antegrade blood flow to the right lung. In cardiac catheterization, the right PA appeared dysplastic compared with the left. Hemodynamic analysis by right ventriculography pictured a wash-in of contrast medium to the right PA and a diastolic reversed flow to the left PA (Fig. 2, Movie 1). All blood from the right atrium flew into the left PA as seen by right ventriculography. It was proposed that blood flow went from the collateral tract arising from the descending aorta via the right lung to the right PA and eventually refluxed into the left PA.

Cardiac 4D flow MRI performed at a 3T scanner (Discovery 750, GE Healthcare, Waukesha, WI, USA) identified most of the blood from the main PA to flow into the left PA. MR parameters included: imaging volume: 32 × 32 × 22 cm, TR/TE = 6.2/2.0 ms, VENC = 200 cm/s, cardio-respiratory system using pulse meter and respiratory bellow signals was used to minimize motion artifacts from breathing. A total of 10,000 unique projection angles were acquired, resulting in
a scan time of approximately 5 min 9 s with slight variations based on the respiratory pattern. In agreement with the 2D phase contrast method, blood flow measurement by 4D flow MRI (Cardio Flow Design, Tokyo, Japan) (Movie 2) showed flow volumes of main PA: 2132 mL/min, left PA: 25689 mL/min, right PA: −22 mL/min, collaterals originating from the aorta: 96 and 91 mL/min (Fig. 3). Consistent with a larger blood flow in the left PA compared with main PA found on CT and missing accumulation in the right lung by scintigraphy, the detected negative flow values in the right PA indicate reversed flow. Additionally, turbulent or to-and-fro phenomenon due to increased pulmonary vascular resistance of the right lung may contribute to negative flow velocity values.

In patients with structural heart diseases, cardiovascular MRI is nowadays a clinically established method representing the gold standard for non-invasive assessment. 4D flow MRI enables the simultaneous assessment of anatomy and functional hemodynamics within only one single acquisition without the need of contrast material. It furthermore delivers accurate quantitative flow parameters and comprehensive blood flow visualization by use of particle tracing, which may even outperform conventional MRI.

In terms of clinical management, three findings of the presented case are of major interest: (1) turbulent flow in the right PA, which may be caused by blood entering the peripheral pulmonary arteries and capillary bed via bronchial arteries, thereby increasing pulmonary arterial pressure, (2) the flow volume to the right lung, as it determines the risk of right pulmonary hemorrhage, and (3) the flow volume to the left lung, as high flow may lead to pulmonary hypertension in the future. With 4D flow MRI we were able to simply quantify these values, which would have been difficult to assess otherwise. The detected lower flow volume in the main PA compared to the left PA supports the finding of collateral flow from the aorta to the right lung. The observed reversed flow in the right PA may be interpreted as to-and-fro phenomenon (based on an increased pressure of the right PA due to the PV atresia and potentially due to bronchial artery-to-PA fistula) or indicate the functioning of the right PA as drainage vessel, as support by the angiogram. That also implies, that the blood flow to the right lung may be estimated by the difference of blood flow between the main and left PA. Referring to recent literature, normal lungs show a left PA:right PA blood flow ratio of 47:53. However, in our case we could not directly observe flow from the peripheral pulmonary arteries into the right PA by CT or 4D flow MRI, reflecting venous return to systemic circulation. The detection of excessive left pulmonary blood flow in our patient, is of high interest for clinical management to monitor the progress of pulmonary hypertension. Collateral vessels from the aorta providing arterial flow to the lung are also observed in patients with bronchopulmonary sequestration, a rare congenital abnormality with nonfunctioning lung tissue lacking connection to the tracheobronchial tree. In our patient, however, the bronchus of the right lung was connected to the trachea.

Fig. 1 Axial contrast-enhanced computed tomography (CT), 3D CT angiography and static pulmonary scintigraphy with $^{99m}$Tc-macro-aggregated albumin. CT images and CT angiography show missing right pulmonary vein. Pulmonary scintigraphy indicates no blood supply to the right lung via pulmonary artery. Note the low density structure in the right hilar region, which most likely correspond to lymph nodes.

Fig. 2 Right ventriculography. Right pulmonary artery presents dysplastic compared with the left. Although the right pulmonary artery (PA) is temporarily visualized, all contrast media in the right PA flows to the left PA. (The movie is available online.)
By enabling comprehensive visualization of blood flow direction and quantification, 4D flow analysis proves to be a valuable tool for clinical work and holds great potential as alternative to routine MRI or invasive catheterization regarding the evaluation and management of complex congenital cardiac anomalies.

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Conflicts of Interest
GE Healthcare and Bayer Healthcare support research projects of U. Motosugi, which are not related to this study. The other authors declare no conflict of interest.

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