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Abstract

We investigated the usefulness of helical computed tomography(CT)in the morphological diagnosis of pulmonary vein stenosis, particularly that in infants and small children. In total, 20 helical CT examinations were performed in 10 post-operative cases of Total Anomalous Pulmonary Venous Drainage(TAPVD), 3 cases of single right ventricle, and 1 case of single left ventricle. In all cases, distinct morphological imaging was possible. Pulmonary vein stenosis could be categorized into three types: (1)stenosis from the anastomosis of the common pulmonary vein (CPV)-the left atrium (LA) to the peripheral pulmonary vein; (2) stenosis only at the anastomosis of CPV-LA; and (3) stenosis due to compression by nearby organs. Coronal views by multiplanar reconstruction (MPR) provided morphological information along the up-down direction of the body axis. Morphological diagnosis of pulmonary vein stenosis is important in deciding prognosis and therapeutic regimens, and helical CT was considered useful for such diagnosis in our 14 young patients.

KEYWORDS: pulmonary vein stenosis, helical CT

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Original Article

Usefulness of Helical Computed Tomography in Diagnosing Pulmonary Vein Stenosis in Infants

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We investigated the usefulness of helical computed tomography (CT) in the morphological diagnosis of pulmonary vein stenosis, particularly that in infants and small children. In total, 20 helical CT examinations were performed in 10 post-operative cases of Total Anomalous Pulmonary Venous Drainage (TAPVD), 3 cases of single right ventricle, and 1 case of single left ventricle. In all cases, distinct morphological imaging was possible. Pulmonary vein stenosis could be categorized into three types: (1) stenosis from the anastomosis of the common pulmonary vein (CPV)-the left atrium (LA) to the peripheral pulmonary vein; (2) stenosis only at the anastomosis of CPV-LA; and (3) stenosis due to compression by nearby organs. Coronal views by multiplanar reconstruction (MPR) provided morphological information along the up-down direction of the body axis. Morphological diagnosis of pulmonary vein stenosis is important in deciding prognosis and therapeutic regimens, and helical CT was considered useful for such diagnosis in our 14 young patients.

Key words: pulmonary vein stenosis, helical CT

There is a possibility that pulmonary vein stenosis advances rapidly in its symptoms, and early diagnosis and appropriate surgical repair are required. To this aim, it is indispensable to grasp the location, extent, and morphology of the stenosis. To date, echocardiography and cardiac catheterization have been the main tools of diagnosis, but they sometimes fail to obtain satisfactory results for several reasons. To achieve more accurate diagnosis, we investigated the usefulness of helical computed tomography (CT) in the morphological diagnosis of pulmonary vein stenosis.

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Materials and Methods

The subjects comprised 14 cases diagnosed as pulmonary vein stenosis by clinical symptoms and examinations such as chest-X-ray and echocardiography. A total of 20 examinations was carried out between March 1998 and April 2004. Ages at examination ranged from one to 64 months, and body weights ranged from 3.2 to 25 kg. The breakdown of the diseases was: 10 post-operative cases of Total Anomalous Pulmonary Venous Drainage (TAPVD) (3 of which concurrently had asplenia), 3 cases of single right ventricle, and 1 case of single left ventricle. The details of the cases are shown in Table 1.

Helical CT equipment used included the single-slice CT Hi Speed Advantage (General Electric Co., Fairfield, CONN, USA), and the multi-slice CT Aquilion, (Toshiba Co., Ootawara, Japan). The CT scanning was

Table 1

Case	Diagnosis	Operation	Age at Helical CT
1	Asplenia, TAPVD(I), SRV, PS, CAVC	TAPVD repair, PAB, PVO release	1 m, 4 m
2	Asplenia, TAPVD(III), SRV, PS, CAVC	TAPVD repair, RV-PA shunt	2 m
3	Asplenia, TAPVD(III), SRV, PS, CAVC	TAPVD repair, Rt.modified BT-shunt	1 m
4	TAPVD(I)	TAPVD repair, PVO release	6 m, 8 m
5	TAPVD(I)	TAPVD repair, PVO release	1 m, 3 m
6	TAPVD(II)	TAPVD repair, PVO release	1 m, 3 m, 6 m
7	TAPVD(I)	TAPVD repair	6 m
8	TAPVD(III)	TAPVD repair	2 y 7 m, 5 y 4 m
9	TAPVD(III)	TAPVD repair	3 m
10	TAPVD(III)	TAPVD repair	3 m
11	DIRV, SRV, severe PS	Rt.modified BT-shunt	10 m
12	DORV, SRV, CAVC, hypo LV, CoA	CoA repair	2 m
13	DORV, SRV, CAVC	PAB	3 m
14	DILV, SLV, straddling MV	PAB, BDG	4 y 9 m

TAPVD, Total anomalous pulumonary venous drainage (I) supracardiac type (II) paracardiac type (III) infracardiac type; SRV, Single right ventricle; PS, Pulmonary stenosis; CAVC, Common atrioventricular canal; PAB, Pulmonary artery banding operation; PVO, Pulmonary vein obstruction; DIRV, Double-innlet right ventricle; BT-shunt, Blalock taussig shunt; DORV, Doubel-outlet right ventricle; LV, Left ventricle; CoA, Coarctation of the aorta; SLV, Single left ventricle; MV, Mitral valve; BDG, Bilataral bidirectional glenn operation.

performed without ECG gating or respiratory arrest. Nonionic contrast medium was used after 1.7- to 2.5-fold (two-fold on average) dilution (Iopamidol, 300 mOsm). Contrast medium was injected at 1.2–2.0 ml/kg (1.6 ml/kg on average) at a speed of 0.6–1.5 ml/sec (1.0 ml/sec on average). The slice thickness was set at 1 mm or 3 mm for single-slice CT. In newborn babies and infants, the images were obtained first with the slice width of 3 mm, and the targeted area was re-scanned with the slice width of 1 mm. In multi-slice CT, the slice width was set at 1 mm uniformly.

For sedation, sodium trichloroethyl phosphate was given orally (80 mg/kg) and secobarbital sodium was infused intravenously (4 mg/kg) when necessary. In no cases did breathing stop during the scanning.

Table 2

Case	Morphological diagnosis of pulmonary vein stenosis	
	Echocardiography	Helical CT
1	×	○
2	×	○
3	×	○
4	×	○
5	×	○
6	×	○
7	×	○
8	×	○
9	○	○
10	×	○
11	×	○
12	○	○
13	×	○
14	○	○

○, morphological diagnosis was possible; ×, impossible.

Results

In all cases, stenosis of the pulmonary vein was identified (Table 2). In 6 (cases 1–6) of the 10 postoperative TAPVD cases, including the 3 cases of asplenia, pulmonary vein stenosis was observed at the anastomosis and its proximity. Even roughly 1 month after the operation for the stenosis in these 6 cases, pulmonary vein stenosis extended to the periphery.

Case 1 had an anastomotic operation of the common pulmonary vein (CPV) and the left atrium (LA); echocardiography measured the velocity of blood influx to the left atrium from the right pulmonary vein at 1.2 m/s on postoperative day 5, but this increased to 2.5 m/s on postoperative day 40. Helical CT on postoperative day 48 clearly visualized the pulmonary vein and stenosis at the opening of the right pulmonary vein (Fig. 1). Since it turned out that the pulmonary vein had a wide enough caliber on the distal side of the stenosis, an operation to release the stenosis was indicated. An operation for the stenosis using the sutureless *in situ* pericardial repair

method was performed 89 days after the first operation. Echocardiography immediately after the operation revealed an improvement in the blood flow velocity to 1.0 m/s at the stenosis.

However, 30 days after the operation for the stenosis, the blood flow increased to 2.2 m/s, and turbulent flow was observed in the left pulmonary vein. Helical CT on postoperative day 41 demonstrated stenosis at and near the opening of the left pulmonary vein that had not been observed previously, and the right pulmonary vein not only had re-stenosis but the stenosis had extended to the periphery (Fig. 2).

In the remaining 4 post-operative cases of TAPVD (cases 7–10), stenosis was found only at the anastomosis.

Case 8 was TAPVD Type 3, and stenosis at the anastomosis became distinct 5 years and 4 months after the anastomosis of CPV-LA. Two years and 8 months after the operation, helical CT showed no stenosis at the

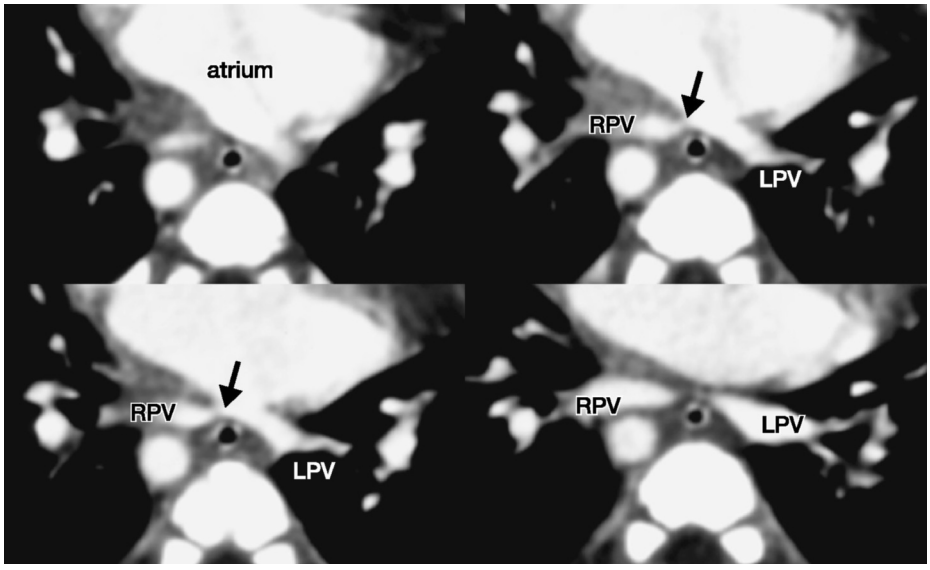

Fig. 1 LPV, Left Pulmonary Vein; RPV, Right Pulmonary Vein; ↓, Pulmonary vein stenosis.

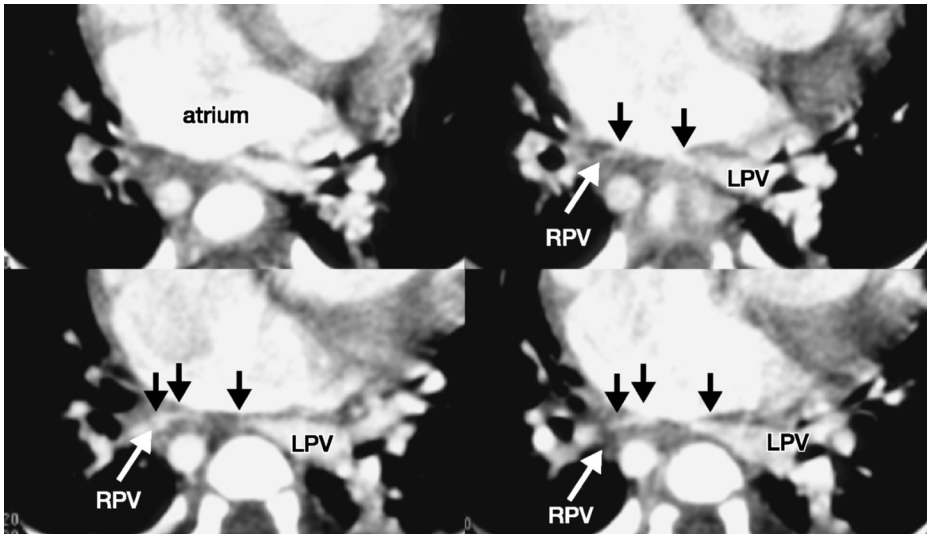

Fig. 2 LPV, Left Pulmonary Vein; RPV, Right Pulmonary Vein; ↓, Pulmonary vein stenosis.

anastomosis (Fig. 3), and cardiac catheterization at the same period revealed no stenosis, either.

Five years and 4 months after the operation, helical CT showed stenosis only at the anastomosis, but not in the adjacent pulmonary vein (Fig. 4). To obtain information along the body axis, images were subjected to multiplanar reconstruction (MPR), and coronal views were reconstructed (Fig. 5). In these cross-sectional images, there was no stenosis in the proximity of the anastomosis in the pulmonary vein, and stenosis limited to the anastomosis was confirmed more precisely. These findings were consistent with those during the operation for stenosis performed later.

Four cases with single ventricle (cases 11–14) had moderate to severe atrial-ventricular (AV) regurgitation during the observation period. Chest X-rays revealed cardiomegaly with cardiac-thoracic ratios (CTRs) greater than 0.60, and shadows of pulmonary vasculature decreased in the left lower lobe.

Case 11 had a single right ventricle, double-inlet right ventricle, and severe pulmonary artery stenosis, and underwent a right modified Blalock Taussig-shunt operation on postnatal day 20. Moderate tricuspid valve regurgitation was observed and CTR was 0.60. During the cardiovascular catheterization 10 months after the operation, pulmonary arteriography revealed insufficient opacification of the left lower pulmonary artery, and contrast medium tended to stay. Helical CT at the same period demonstrated that the left lower pulmonary vein was compressed from the front by the atrium and from the back by the descending aorta and the spine (Fig. 6).

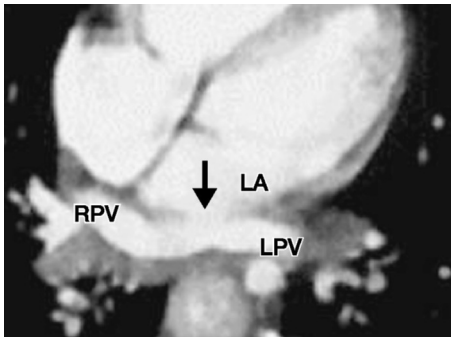

Fig. 3 LA, Left Atrium; LPV, Left Pulmonary Vein; RPV, Right Pulmonary Vein; ↓, anastomosisi of common pulmonary vein-left atrium.

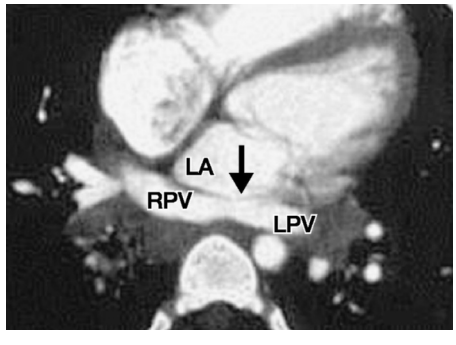

Fig. 4 LA, Left Atrium; LPV, Left Pulmonary Vein; RPV, Right Pulmonary Vein; ↓, stenosis at the anastomosisi of common pulmonary vein-left atrium.

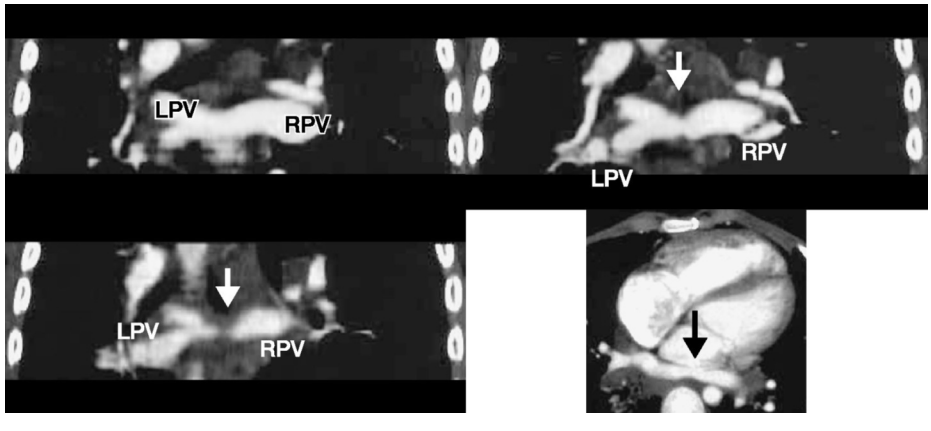

Fig. 5 LPV, Left Pulmonary Vein; RPV, Right Pulmonary Vein; ↓, stenosis at the anastomosisi of common pulmonary vein-left atrium.

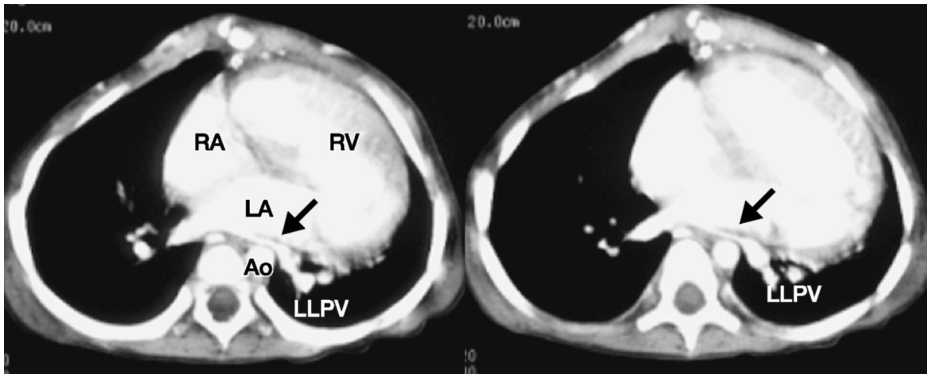

Fig. 6 Ao, descending Aorta; LA, Left Atrium; LLPV, Left Lower Pulmonary Vein; RA, Right Atrium; RV, Right Ventricle; ↓, left lower pulmonary vein was compressed by LA and Ao.

Discussion

It is possible to evaluate the presence or absence of pulmonary vein stenosis by echocardiography. Especially, measurement by pulse and continuous wave Doppler of the velocity over time at the pulmonary vein stenosis is useful for evaluating the advance of pulmonary vein stenosis [1].

However, as shown in cases 1–3, in which blood flow to the lungs and to the body were provided by the same single ventricle, an advance of stenosis may not be consistent with a change in velocity. This may be because an advance of stenosis increases pulmonary vessel resistance and decreases pulmonary blood flow. Indeed, in our experience, velocity at the stenosis ranged from 2.0 to 2.5 m/s. It is difficult to elucidate the morphology of the pulmonary vein in the extra-mediastinum and the lung by echocardiography. Furthermore, we sometimes experience difficulty in clearly visualizing even the shape of the pulmonary vein near the anastomosis of CPV-LA. In color Doppler examination, adaptation to decrease the flow velocity range is necessary because the blood flow is slow proximal to the stenosis, and clear visualization is difficult (Table 2).

To compensate for these disadvantages and evaluate the morphology of the stenosis, cardiovascular catheterization is often carried out [2, 3]. However, this method has serious defects in terms of the clarity of the images and the difficulty of grasping the positional relations to the surrounding organs. In addition, it is not rare for the patients' conditions to be aggravated after such examination.

Magnetic resonance image (MRI) is a very useful modality because it can provide clear images and information on blood flow [4–6]. However, it is difficult to examine patients with dyspnea and critical general conditions because imaging requires a long period of time.

As presented herein, helical CT should be considered suitable for infants and newborn babies with complicated heart anomalies [7, 8] who are difficult to keep still, because it takes only a short period of time [3, 4]. Recently, the introduction of multi-slice CT [9–11] with advances in speed has made the examination even easier. The volume of contrast medium used in this study was 1.6 ml/kg on average, and the examination was less invasive than cardiovascular catheterization, resulting in no case with aggravated conditions after the examination.

In our study, helical CT provided clear images of the shape of the pulmonary vein, and made it possible to accurately evaluate the morphology and extent of pulmonary vein stenosis. As has been said, it was easy to grasp the positional relations between the pulmonary vein and the surrounding organs [12]. In this study, pulmonary vein stenosis could be categorized into 3 types: (1) stenosis from the anastomosis of CPV-LA to the peripheral pulmonary vein; (2) stenosis only at the anastomosis of CPV-LA; and (3) stenosis due to the compression by surrounding organs. It was presumed that more accurate visualization was possible without ECG gating and respiratory arrest because vasculature including the pulmonary vein is hardly moved by the pulse of the heart, and adhesion is present, especially in postoperative cases.

It is possible to prepare three-dimensional images with relative ease by helical CT, because the data has three-dimensional continuity [13, 14]. We presented MPR that could provide sectional images arbitrarily. As shown

in the images, image reconstruction is possible not only in the front-back direction in relation to the body axis, but also in the up-down direction. Helical CT image data in three directions are called isotropic voxel data, and use of these data is expected to reduce stair-step artifacts and increase diagnostic efficacy [15]. It is also possible to evaluate stenosis from different angles and diagnose the morphology more accurately. There are SSDs (Shaded Surface Displays), MIPs (Maximum Intensity Projection [s]), and other three-dimensional volume rendering methods in helical CT; it is necessary to choose and use them with understanding of their various advantages and disadvantages.

Surgical repair [16–18], balloon dilatation [19, 20], and stent implanting [17, 21, 22] have been the options in treating pulmonary vein stenosis. There is a possibility of re-stenosis in any treatment option, but, to date, the first indication would be the surgical release of stenosis. Planning the operation requires detailed information on the stenotic lesion; the location, extent, morphology, and cause of the stenosis need to be examined thoroughly, and we must judge whether a general operation for stenosis is sufficient or the sutureless *in situ* pericardial repair method is necessary. When pulmonary vein stenosis is extended to the periphery distally to the anastomosis, the case is resistant to treatment and the prognosis is extremely poor. From this point of view, helical CT was able to provide highly accurate information compared with other modalities and was useful in deciding the treatment strategies for our 14 young patients.

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