Total Anomalous Pulmonary Venous Connection: Helical Computed Tomography as an Alternative to Angiography

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Background: Echocardiographic evaluation of the pulmonary veins is inadequate at times. Cardiac catheterization, especially in sick neonates, may be a high-risk procedure. Helical computed tomography with three-dimensional reconstruction is noninvasive but remains an underutilized modality.

Methods and Results: Between January 2002 and February 2003, 7 computed tomography scans of children 3 weeks to 5 years of age were performed to evaluate the drainage of the pulmonary veins in suspected total anomalous venous drainage. Helical computed tomography (GE High speed Advantage) was performed using 2 mm sections, and rapid bolus hand injections of 2 ml/kg body-weight of nonionic intravenous contrast. Sagittal and coronal reformats, and three-dimensional reconstructions were performed, and reviewed by the radiologist. The findings were discussed with the pediatric cardiologist and surgeon involved in the case. The diagnoses included complex congenital heart disease (n=5), isolated infradiaphragmatic total anomalous pulmonary venous connections (n=1), and transposition of the great arteries with total anomalous pulmonary venous connections (n=1). Cardiac computed tomography accurately demonstrated infradiaphragmatic total anomalous pulmonary venous connections in 4, and supracardiac drainage in 3 patients, in addition to the other cardiac findings. The findings on computed tomography scan correlated with surgical (n=5) and/or angiographic findings (n=2) in 7 patients.

Conclusions: In sick, high-risk patients, cardiac computed tomography can be considered as an alternative to cardiac catheterization for the evaluation of pulmonary venous drainage. **(Indian Heart J 2003; 55:624–627)**

Key Words: Congenital heart disease, Total anomalous pulmonary venous connections, Helical computed tomography

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Total anomalous pulmonary venous connections (TAPVC) are generally easily diagnosed on echocardiography. However, the exact site of drainage of the pulmonary veins may occasionally be in doubt, especially in association with complex cardiac disorders. Although angiography has been the gold standard for evaluation, it carries certain inherent risks, especially in small and sick infants. Computed tomography (CT) is noninvasive, and has a lower incidence of complications. However, reports of its use in the assessment of TAPVC are limited. We prospectively used helical CT to assess abnormalities of the pulmonary veins diagnosed or suspected by echocardiography in children with complex congenital heart disease. CT findings were correlated with echocardiographic, angiographic, and/or surgical findings.

Methods

Between January 2002 and February 2003, 7 CT scans were performed to evaluate the drainage of the pulmonary veins in children with suspected TAPVC, aged 3 weeks to 5 years. Informed consent was taken from one of the parents of the child. Helical CT (GE High speed Advantage) was performed using 2 mm sections at a pitch of 1:1, and intravenous rapid bolus hand injections of 2 ml/kg bodyweight of nonionic contrast. A delay of 30 s was given between the beginning of the contrast injection and the start of the scan. Each patient was scanned from the apex of the lung to the iliac crests. To avoid motion artifact, children were sedated as required. Sagittal and coronal MPR reformats, and three-dimensional (3-D) MIP reconstructions were obtained. A cardiac radiologist, who was experienced in treating congenital heart diseases and blinded to the echocardiographic diagnosis, reviewed the cardiac CT scans. After a diagnosis was arrived at, the findings were discussed with the pediatric cardiologist and surgeon involved in the case.

Results

The details and diagnoses of the 7 patients with TAPVC who were studied are given in Table 1. The diagnoses included complex congenital heart disease (*n*=5), isolated infradiaphragmatic TAPVC (*n*=1), and transposition of the great arteries with TAPVC (*n*=1). Cardiac CT scan accurately demonstrated supracardiac drainage in 3 patients (Fig. 1), and infradiaphragmatic TAPVC (Figs 2 and 3) in 4 patients in addition to other cardiac findings. In 7 patients, the findings on CT scan correlated with surgical (*n*=5) and/or angiographic findings (*n*=2). Obstruction was suggested on the basis of anatomic narrowing of the pulmonary veins noted on CT scan.

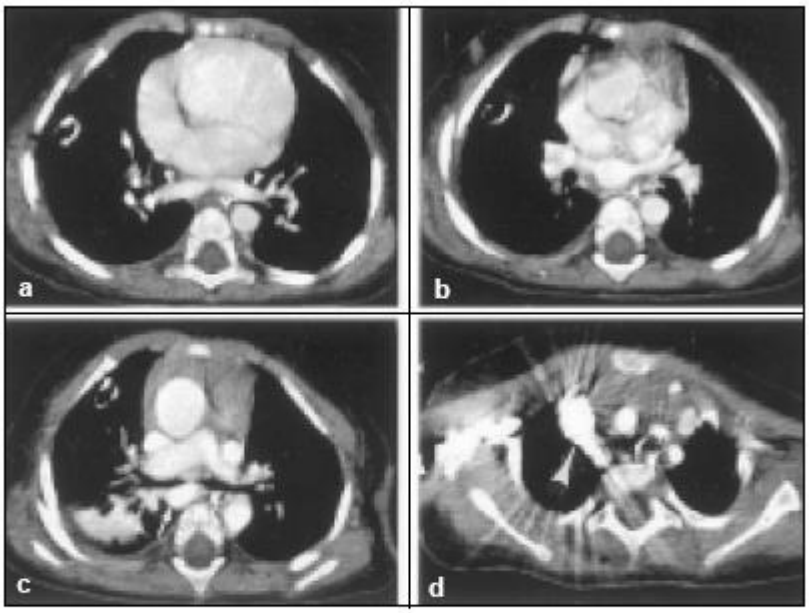

Fig. 1. *Supracardiac TAPVC draining into the superior vena cava.* (**a**) *Right and left inferior pulmonary veins (short arrowheads) joining posterior to the left atrium into a common chamber.* (**b**) *Right and left superior pulmonary veins draining into the common chamber (open arrowhead).* (**c**) *Vertical vein (short arrow) ascending posterior to the right main stem bronchus.* (**d**) *Vertical vein (long arrowhead) draining superiorly into the superior vena cava.*

Table 1. Details of patients and diagnoses

Age	Sex	Type of TAPVC	Connection	CT evidence to suggest obstruction	Associated anomalies
3 months	M	Infracardiac	Middle hepatic vein	Yes	Situs ambiguous, dextrocardia, single ventricle, d-malposed arteries, pulmonary hypertension
5 years	M	Supracardiac	SVC	No	Transposition of the great arteries
5 months	M	Infracardiac	Portal vein	Yes	Unbalanced atrioventricular canal, pulmonary atresia VSD, malposed great vessels, PDA
3 weeks	M	Infracardiac	Portal vein	Yes	–
10 months	M	Supracardiac	Right SVC	No	Situs inversus, asplenia, atrioventricular canal, malposed great arteries, pulmonary atresia
2 months	M	Infracardiac	Portal vein	Yes	Situs inversus, asplenia, double-outlet right ventricle pulmonary atresia, PDA
2 years	F	Supracardiac	Right SVC	No	Situs inversus, unbalanced atrioventricular canal, d malposed vessels, pulmonary stenosis

M: male; F: female; TAPVC: total anomalous pulmonary venous connection; CT: computed tomography; VSD: ventricular septal defect; PDA: patent ductu arteriosus; SVC: superior vena cava

Discussion

According to Darling et al.,[1] there are 4 types of TAPVC: supracardiac, cardiac, infracardiac, and the mixed variety.[2] In most cases, a common chamber is seen posterior to the left atrium. In complex congenital heart disease, where pulmonary blood flow is low, the common chamber is also correspondingly diminutive and easily missed on echocardiography.

In cases in which echocardiography is equivocal, angiography has been used to delineate the pulmonary venous drainage. However, angiography has certain inherent risks and, at times, cannot be performed. Additionally, in complex congenital heart diseases, accurate visualization of the pulmonary venous anatomy may not be obtained even after angiography. During cardiac catheterization in these patients, entry into the pulmonary artery may at times be dangerous, and sometimes impossible. In such a situation, just a ventricular injection may not reveal any discernible dye in the pulmonary venous system even on prolonged levophase. Therefore, a noninvasive and relatively inexpensive alternative to angiography would be helpful in the evaluation of children with complex congenital heart disease. Although magnetic resonance imaging (MRI) is well described,3–5 it remains expensive, and not all cardiac centers have this facility. CT is much more affordable than MRI and, especially in the Asian context, far more cardiac centers have access to it than to a cardiac MRI.

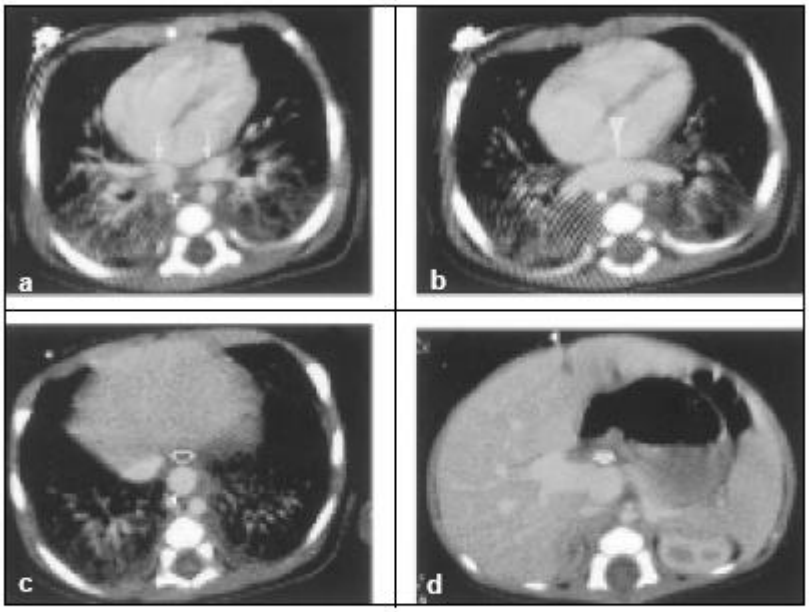

Fig. 2. *Infracardiac TAPVC draining into the portal vein.* (**a**) *Right and left superior pulmonary veins (short arrows) joining a common chamber posterior to the left atrium.* (**b**) *Right and left inferior pulmonary veins also draining into the common chamber (long arrowhead).* (**c**) *Large pulmonary venous channel (open arrowhead) anterior to the descending thoracic aorta.* (**d**) *Pulmonary venous channel (wide arrowhead) draining into the portal vein.*

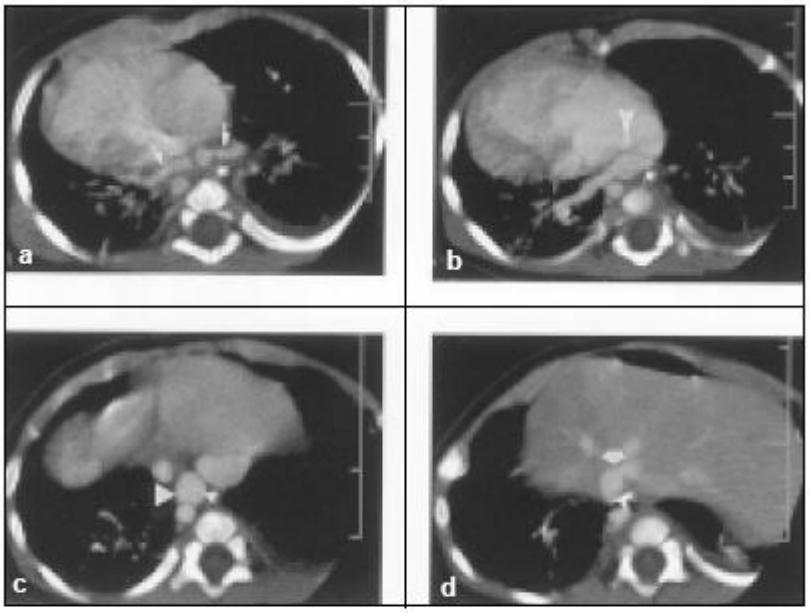

Fig. 3. *Infracardiac TAPVC draining into the middle hepatic vein.* (**a**) *Right and left superior pulmonary veins (short arrows) draining into the common chamber, with associated dextrocardia.* (**b**) *Right and left inferior pulmonary veins also draining into the common chamber (long arrowhead).* (**c**) *Large pulmonary venous channel (short arrowhead) anterior to the right-sided descending aorta.* (**d**) *Channel (wide arrowhead) draining into the middle hepatic vein with narrowing at its distal end.*

Although there are several case reports on the use of helical CT in complex congenital heart diseases in children,[6–11] specific studies evaluating pulmonary venous drainage are limited.[12] Kim et al.,[12] in their study of 14 patients with

TAPVC, demonstrated that the combination of axial and 3-D images in helical CT angiography are helpful in the assessment of TAPVC, and this combination can be a good diagnostic tool for the preoperative evaluation of neonates and infants.

According to Kawano et al.,[13] 3-D helical CT angiography can clearly demonstrate the shape and spatial relation of the great arteries, the proximal branch pulmonary arteries, and anomalous pulmonary venous connections. However, intracardiac structures could not be visualized because of blurred and/or unclear edges of the ventricular wall, caused by respiratory movement.

CT angiography is a noninvasive procedure, which can provide detailed information on the anatomic features and relations of both the great vessels as well as other extracardiac structures that are very important from the perspective of a cardiac surgeon. Helical CT angiography can be performed considerably quicker than cardiac catheterization; thus, general anesthesia can be avoided, which is a great advantage, especially in the case of a sick baby.

Currently, helical CT angiography cannot visualize intracardiac structures well. In addition, the images of extracardiac structures may have blurred edges because of respiratory and cardiac movements. Another limitation of CT angiography is that it cannot provide pressure gradients; the anatomic configuration of the vessels may suggest pressure effects. However, in most cases, echocardiography gives true estimates of pressure gradients. One of the potential limitations of CT is that in the presence of diffusely small pulmonary veins (<2 mm), CT angiography may not clearly demonstrate the course of these small veins.

In our study, CT accurately demonstrated the pulmonary venous drainage in all 7 patients with TAPVC. Thus, computed helical CT angiography with 3-D reconstructions can be considered a safe and noninvasive alternative to cardiac catheterization in the evaluation of TAPVC, especially in the setting of complex congenital heart disease.

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