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The Role of Echocardiography in Diagnosing Crossed Pulmonary Arteries

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ABSTRACT

Background: Crossed pulmonary artery (CPA) is a rare anomalies and it does not cause significant hemodynamic repercussions, but still has clinic significance. The diagnosis of CPA is more relay on computer tomography (CT) or cardiac angiography. We aimed to analysis the value of transthoratic echocardiography in the diagnosis of CPA to see whether it can be diagnosis by echocardiography accurately.

Method: Retrospective analysis the echocardiographic data of CPA in our single-center from January 2014 to December 2016. The Vivid 7 Dimension (GE) with M4S transducer, Vivid i Cardiac system (GE) with 7S-Rs transducer and iE33 Ultrasound (Philips) with S8-3 and X5-1 transducer with a frequency of 5 to 7.5 MHz were used.

Result: Totally twenty-nine patients were diagnosed as CPA by echocardiography. Except tow cases were misdiagnosed as pulmonary artery sling (PAS) at first time, the other patients got definite diagnosis by echocardiography. Seven patients had MSCT and three patients underwent surgery for other intracardiac malformations, which were consistent with the echocardiography. Sixteen patients with other intracardiac malformations, including eight atrial septal defect, four ventricular septal defect, one double outlet of right ventricular, one triatriatum, one patent ductus arteriosus and one bicuspid aortic valve. Thirteen single CPA including one patient was Kawasaki disease with coronary aneurysm - like dilatation. Sixteen patients coexist with mild stenosis of the pulmonary branches.

Conclusion: Echocardiography is the preferred method of CPA and can make definite diagnosis of it, which should be made differential diagnosis with PAS. Sweep superior-inferior at the high left parasternal view of the bifurcation of the pulmonary artery is important to diagnose CPA.

Keywords: Echocardiography, Crossed pulmonary artery, Pulmonary artery sling, Pulmonary bifurcation

INTRODUCTION

Crossed pulmonary artery (CPA) is a rare but cannot be ignored form of malposition of the branch pulmonary arteries [1], which means both branches of pulmonary artery cross each other on their course proceeding to the corresponding lung, thus forming a crisscross pattern [2]. According to the degree of its cross, it could be classified into two types, the typical and the atypical. Single CPA usually had no specific clinical manifestations. However, CPA often coexists with other intracardiac or extracardiac anomalies, and also chromosomal abnormalities. Therefore, further evaluation may be required to manage these patients with CPA. In some cases, a multidisciplinary consultation should be carried out [3].

In previous reports, CPA was generally diagnosed by computer tomography or cardiac angiography. No one applies for echocardiography because of suspecting CPA, but echocardiologists should be acquainted with CPA to avoid misdiagnosing it as other pulmonary vascular malformations, such as pulmonary artery sling (PAS). To explore the possibility of diagnosing CPA by using echocardiography, we retrospectively analyzed our imaging data of CPA.

METHODS

Study population

The retrospective study was approved by our institutional research ethics board, and the need for patient consent was waived. All children diagnosed with CPA by echocardiography between January 2014 and December 2016 was identified from the electronic database of Children's Hospital of Fudan University.

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DATA COLLECTION

Electronic charts of the children with CPA were reviewed. Presentation of symptoms and echocardiographic parameters of patients were collected. A form was created to collect the presentation symptoms of the patients, including murmurs, shortness of breath, or no symptoms. Every patient was examined by echocardiography during their first admission in our hospital and concomitant cardiovascular anomalies were recorded. All echocardiographic data were carefully analyzed.

STUDY DESIGN

The initial seven patients who were suspected as CPA by echocardiography were transferred to multi-slice spiral computer tomography (MSCT) exam. We compared the echocardiographic images with the MSCT images retrospectively to analyze the morphology of the pulmonary branches. The other twenty-tow cases were diagnosed only by echocardiography.

Echocardiographic Imaging

Two-dimensional and Doppler echocardiography were performed with GE Vivid 7 Dimension (GE-Vingmed Ultrasound AS, Horten, Norway) with M4S transducer, GE Vivid i Cardiac system (GE Medical system Israel) with 7S-Rs transducer and iE33 imaging system (Philips Healthcare, Bothell, Eberett Highway, Washington, USA) with S8-3 and X5-1 transducer with a frequency of 5 to 7.5 MHz. The left lateral decubitus and supine position was taken and, if necessary, 10% chloral hydrate was administered at a dose of 0.1 mg/kg for sedation. Echocardiographic images were obtained in the parasternal, apical and sub-xiphoid views together with color Doppler echocardiography.

The high left parasternal view and the supra-sternal fossa view were emphasized to observe the pulmonary bifurcation. The left parasternal long-axis view was adopted to display the abnormal branch arising from the ascending aorta. The supra-sternal fossia view to display the anatomical structure of the aortic arch as much as possible to illustrate the position, development, and other abnormalities of the aortic arch. To clarify the origin, the flow of the pulmonary branch was shown by color Doppler. The velocity of the pulmonary branches was measured by pulse-wave or continuous-wave Doppler to see if narrow or not.

MSCT imaging

MSCT was performed in the initial seven patients. Three-dimensional reconstruction was done to show more details about the spatial structure of the pulmonary branches. The relationship of the pulmonary vessels and trachea was also demonstrated. Other co-exited anomalies were diagnosed simultaneously.

STATISTICAL ANALYSIS

Data were presented as frequencies (percentages), medians (ranges) or means ± standard deviations. Differences between groups were tested with the χ^2 test. P value < 0.05 is considered as significant difference. Data analysis was carried out using SPSS version 16.0.

RESULTS

Clinical characteristics of CPA

Between January 2014 and December 2016, 29 patients (17 males and 12 females; age range: 4 days to 10 years at the time of initial diagnosis; median age: 5 months old) who were diagnosed as CPA by echocardiography enrolled in our study.

The complains of patients with CPA include heart murmurs (72.4%, n=21), mostly was a mild murmur at the left sternum border, health examination (17.2%, n=5), shortness of breath or other respiratory symptoms (6.9%,n=2) , and other diseases needed echocardiographic examination, such as Kawasaki disease(3.5%,n=1).

Sixteen patients coexisted with other congenital intracardiac malformations, including atrial septal defect (27.5%,n=8), perimembrane ventricular septal defect (13.8%,n=4), double outlet of right ventricular (3.5%,n=1), triatriatum (3.5%,n=1), patent ductus arteriosus (3.5%,n=1) and bicuspid aortic valve (3.5%,n=1). The other thirteen cases were the single CPA with one patient was coronary aneurysm related to Kawasaki disease. **(Table 1).**

Table 1. Clinical characteristics of CPA

Variable	**Total(n=29)**
Median age(months)	5 (range 4 days to10 years old)
Male:Female	17:12
Symptoms	
Precordial murmurs	72.4%(21/29)
Healthy examination	17.2%(5/29)
Respiratory symptoms	6.9%(2/29)
Others	3.5%(1/29)
Concomitant cardiovascular anomalies	58.6%(17/29)
Atrial septal defect	27.5%(8/29)
Ventricular septal defect	13.8%(4/29)

Patent ductus arteriosus	3.5%(1/29)
Double outlet of right ventricular	3.5%(1/29)
Triatriatum	3.5%(1/29)
Bicuspid aortic valve	3.5%(1/29)
Coronary aneurysm related to Kawasaki disease	3.5%(1/29)

Types of CPA and their echocardiographic characteristics

The direct sign of CPA was displayed at the high left parasternal short-axis view and the suprasternal fossa view, while the typical bifurcation disappeared in the routine left parasternal view. For the typical form, moving the probe up 1 or 2 intercostals in the high parasternal view and sweeping from superior to inferior to show the left pulmonary artery was located in the right to the pulmonary artery and above the right pulmonary artery, and the right pulmonary artery was located in the left to the pulmonary artery and below the left pulmonary artery **(Figure 1**). For atypical form, in the high left parasternal view, the main pulmonary artery continues to the right or left pulmonary directly **(Figure 2**), then the left pulmonary artery was located just above the right pulmonary artery or the right pulmonary artery was located below the left pulmonary artery. There were no other indirect echocardiographic signs of the single CPA.

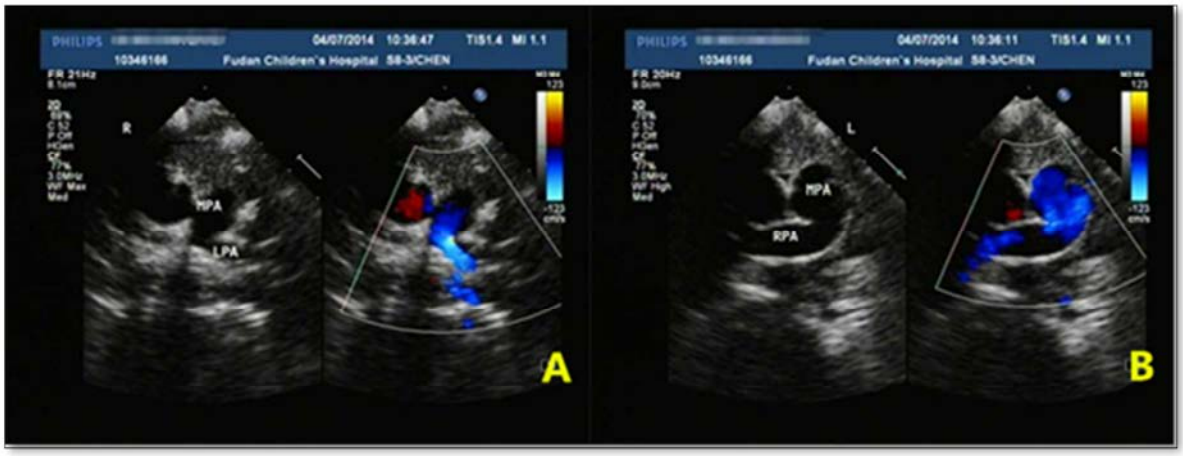

Figure 1. Two-dimensional and Doppler echocardiography from high left parasternal short axis view to show the typical form of CPA: A: the origin of LPA superior and to the right of RPA. B: the origin of RPA inferior to the left of LPA.
MPA=main pulmonary artery, LPA=left pulmonary artery, RPA=right pulmonary artery

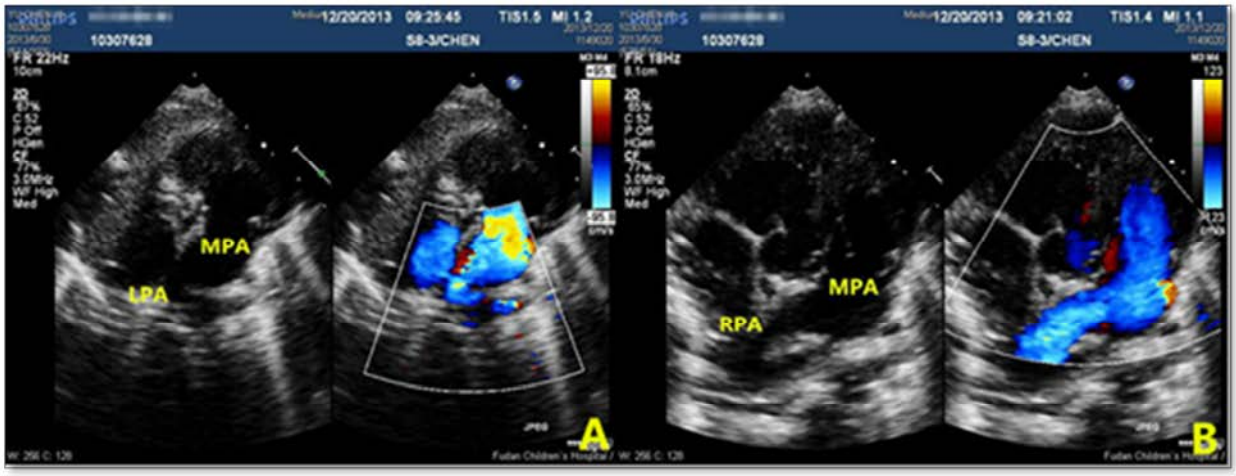

Figure 2. Two-dimensional and Doppler echocardiography from high left parasternal short axis view to show the atypical form of CPA: A: the origin of LPA superior and to the right of RPA. B: RPA was located at its normal place.
MPA=main pulmonary artery, LPA=left pulmonary artery, RPA=right pulmonary artery

The initial diagnose rate was 89.7% (26/29). Two cases were diagnosed as PAS initially, and corrected as CPA by repeat echocardiography. One case showed unclear origin of the left pulmonary artery by bedside echocardiography, then confirmed as CPA by follow-up echocardiography.

Two cases with the typical form while twenty-seven cases with the atypical form of CPA, among which twenty-six cases with left pulmonary artery cross and the another one case with right pulmonary artery cross.

Totally, sixteen cases of CPA were complicated by the stenosis of the pulmonary branches, including twelve cases with left pulmonary artery stenosis (mean velocity 2.44 ± 0.38(m/s), ranged from 2.08m/s to 3.34m/s), one case with right pulmonary artery stenosis (velocity 3.57m/s), and three cases with mild stenosis at both branches (mean velocity 2.77 ± 0.67(m/s) and 2.37 ± 0.33(m/s), ranged from 2.33m/s to 3.54m/s and 2.05m/s to 2.7m/s, respectively). (**Table 2).**

Table 2. Specific Echocardiographic Findings

Echocardiographic Findings	**Total(n=29)**
Types	
Typical form	6.9%(2/29)
Atypical form	93.1%(27/29)
LPA cross	89.7%(26/29)
RPA cross	3.5%(1/29)
Stenosis of the pulmonary branches	55.2%(16/29)
LPA stenosis	41.4%(12/29)
RPA stenosis	3.5%(1/29)
Both branches stenosis	10.3%(3/29)

CPA in MSCT and intraoperative diagnosis compared to echocardiography

MSCT was performed in the initial seven cases. Among these patients, one patient with unclear origin of the left pulmonary artery and two patients diagnosed as PAS at first time, which were diagnose as CPA by the follow-up echocardiography. Left pulmonary artery at the right of the pulmonary artery appeared in the imaging firstly in a set of sequential scanning imaging, and then came with the right pulmonary artery, which located in its normal place or at the left of the pulmonary artery (**Figure 3).** That was in accordance with the echocardiaographic imaging. Three-dimensional reconstruction imaging could show it more clear. None of the patients had the vascular ring. Only one patient with right aortic arch and aberrant left subclavian artery was diagnosed by MSCT. There was no statistical difference between the initial echocardiographic diagnosis and MSCT (p=0.096).

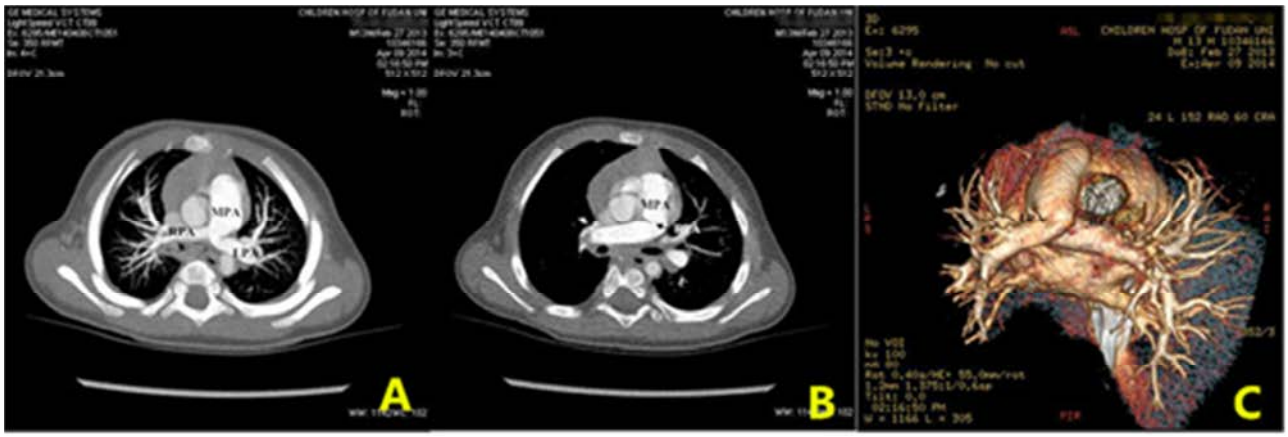

Figure 3. MSCT and 3D reconstruction of the same patient in Figure 1: LPA at the right of the pulmonary artery appeared in the imaging firstly in a set of sequential scanning imaging, and then came with the RPA. It was so similarly to the Figure 1. The 3D reconstruction showed the MPA rotated and the LPA was above the RPA and crossed each other more intuitively.
MPA=main pulmonary artery, LPA=left pulmonary artery, RPA=right pulmonary artery

Three patients with congenital heart diseases received surgery. One case with large ventricular septal defect and two cases with atrial septal defect underwent surgery for repairing the defects and without correcting pulmonary artery branches. All these patients were left pulmonary artery crossed and were confirmed during operation, which show the left pulmonary artery lies above the right pulmonary artery and make a cross with right pulmonary artery (**Figure 4**). The crossed pulmonary was not corrected during surgery.

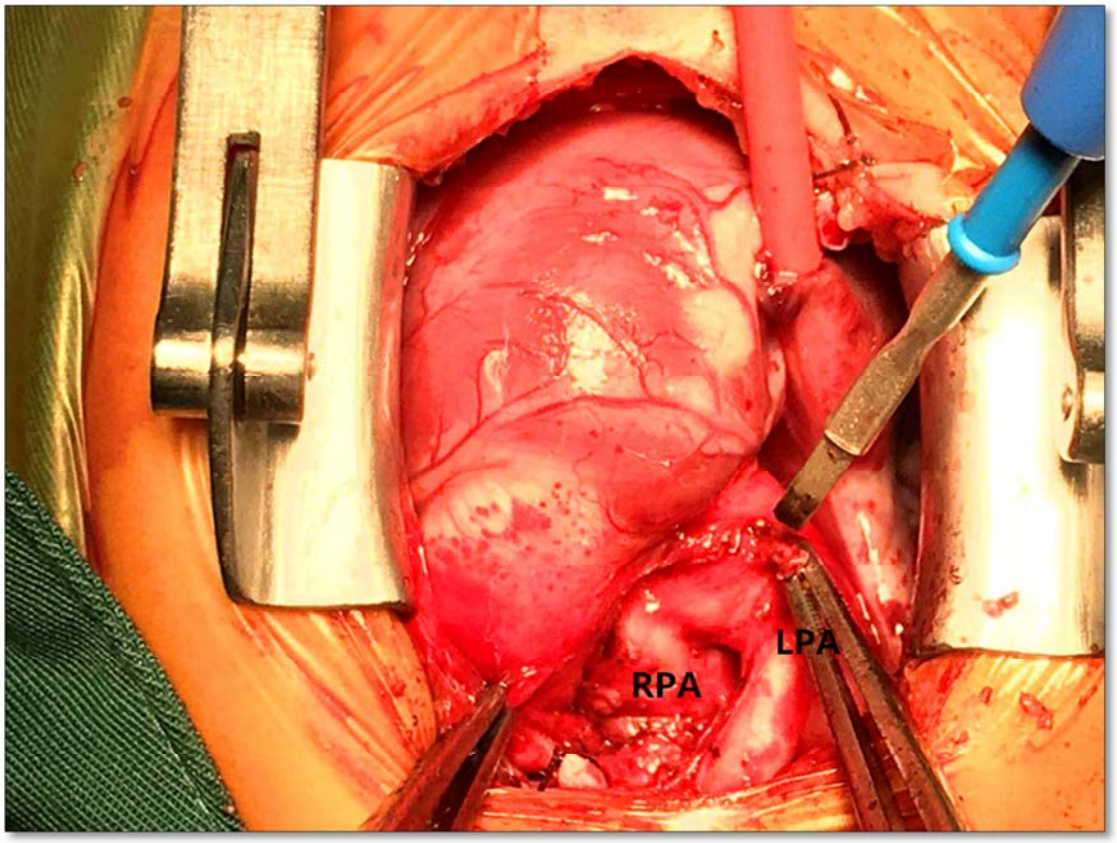

Figure 4. seeing in intraoperative of repairing the ventricular septal defects: the surgeon unfolded the original of the LPA and RPA to show that the ostium of LPA superior and to the right of RPA.
LPA=left pulmonary artery, RPA=right pulmonary artery

The remaining cases were recommended for long-term follow-up by echocardiography. Up to now, no patients with branch pulmonary artery stenosis got worse.

DISCUSSION

In 1966, Jue et al first reported CPA [4], the classic form also called typical CPA was the pulmonary branches cross each other, the ostium of the left pulmonary artery originated superior to the right pulmonary artery and to its right, and the right pulmonary artery originated inferior to the left pulmonary artery and to its left. Thus, the two branches crisscross one another along their courses to their related lungs. In 1970, Becker et al reported the less form also called atypical CPA [5], that is left pulmonary artery openings directly above the right pulmonary artery walking into the left lung without crossing, and this type was more common, while the right pulmonary artery openings directly below the LPA is rare. Its pathogenesis is still unclear. It may be associated with embryonic pulmonary total counterclockwise rotation or due to faulty differential growth during the partitioning of the truncus arteriosus into the aorta and pulmonary trunk, so CPA is easily associated with intracardiac abnormalies especially the conotruncal and aortic arch defects [6], extracardiac malformations [7] and are associated with chromosome 22q11.2 microdeletion [8,9].

Single CPA was asymptomatic. According to our result, if the branch of the pulmonary artery curvature due to narrow or flow faster, a systolic murmurs could be heard at the second intercostal of left sternal border, but always mild. Most patients were diagnosed by accident when they received echocardiograhic examination for other reasons, such as suspected vascular rings, heart murmurs and health examination.

Some centers suggested that diagnosis of malposition of the

branch pulmonary arteries could be challenging if based solely on echocardiography [3], but we don't think so. MSCT could help us to comprehend the spatial structure of the CPA more directly, but it is radioactive. For the safety of our children, MSCT required indications. In the beginning of our research, we had less experience of this malformation. We suggested MSCT and all of them were confirmed as CPA. The subsequent patients didn't do MSCT unless with double of the other vessels abnormality. The left pulmonary artery appeared at first in a set of the imaging which located at the right of the pulmonary artery. Then, came with the right pulmonary artery. This was in accordance with the echocardiographic imaging. When we swept from superior to inferior at the high left parasternal, we found the left pulmonary artery at first and then the right pulmonary artery. We thought echocardiography could instead of MSCT in diagnosis of the CPA without other vascular malformation by comparing the imaging of them according to our result.

As a convenient, economical and safe imaging technique, echocardiography was the primary method used to make definite diagnosis of CPA in our series. Usually, left parasternal view hardly showed the bifurcation of the pulmonary artery. We must made superointerior sweeps from the high left parasternal view and added color Doppler to confirm this abnormality. By observing this view, always found the left pulmonary artery lying superior and to the right of the main pulmonary artery and extending to the left and the right pulmonary artery was lying inferior from the left extending to the right, which is the typical type. If we found that the left pulmonary artery was lying superior on the right pulmonary artery to the left, which is the atypical type. If we failed to obtain the image of the bifurcation of the pulmonary arteries on echocardiography, it may be a clue of malposition of pulmonary arteries branches. We must sweep at the high left parasternal view from superior to inferior more carefully to find the branches.

Single CPA usually does not affect hemodynamics, so it does not require special intervention. Unless CPA cause stenosis of the branches, it might need treatment according to the degree of stenosis. But the diagnosis of CPA had some other meanings. Abnormalities of pulmonary artery branches include PAS, anomalous origin of one pulmonary artery (AOPA) [10] and CPA. In the echocardiographic diagnosis, if the bifurcation of the pulmonary artery disappeared, we should look for the branches to resolve the question of where were the branches. At the supra-sternal fossia view, we demonstrated the ascending aorta and the aorta arch clearly to see if any of the branches arise from it to exclude AOPA [11]. But AOPA always accompanied with severe pulmonary hypertention, the echocardiography showed dilated right atrium and right ventricular and high pressure gradient of tricuspid valve regurgitation, while CPA is not. On the other hand, CPA was more common to confused with PAS. In our study, two patients were diagnose as PAS at first, which were corrected by the following echocardiagraphy and also confirmed by MSCT as left pulmonary artery crossed. PAS is the left pulmonary artery originated in the right pulmonary artery and bypass the trachea formatting of vascular rings result in airway narrowing [12], which should receive MSCT to access the compression degree and development degree of trachea and it required surgical intervention. Because the left pulmonary artery of CPA was originated at the right of main pulmonary artery, the initial region might be overlap with the initial region of right pulmonary artery and then extended to the left into the left lung, especially for the atypical type. The image may be similar to the left pulmonary artery originated from right pulmonary artery. But CPA itself usually does not form vascular rings, resulting in airway narrowing [3]. In the echocardiography of the identification point was to clearly show the ostium of the left pulmonary artery. The important views of this include the high left parasternal view and supra-sternal fossa view of the pulmonary branches. Color Doppler echocardiography enabled confirmation of blood direction of pulmonary artery branches, as well as assessment of the stenosis of them.

Since 1966, the number of reported CPA in the world no more than 60 cases, which always were reported sporadically [13,14]. Echocardiography is the preferred method to diagnose CPA as a non-invasive examination [15,16]. Since January of 2014, we paid more attention of CPA, and diagnosed 29 cases in three years. We estimated the morbidity of CPA might be more than reported. Missed diagnosis often because of: firstly, echocardiography physician does not pay attention to branch pulmonary arteries of normal children; secondly, notice of other intracardiac malformations, easily overlooked the bifurcation; thirdly, because it does not cause meaningful hemodynamic effects, even if found the CPA not diagnosed. However, the diagnosis of CPA has a certain clinical significance, if echocardiography diagnosed as CPA, children may not need further image examination, such as MSCT, which had radioactivity, unless echocardiography showed unclear of the bifurcation or had other anomalies of the aorta arch, but instead to have comprehensive clinical evaluation and molecular examination according to clinic manifestation. So we should carefully studied morphology of pulmonary artery ostia and branches by echocardiography.

LIMITATIONS

The study samples are small and may have been influenced by the limited cognition of the CPA for every echocardiologist and clinicians in our center. All the patients diagnosed as CPA did not undergo further clinical examination to access the extracadiac and chromosomal abnormality.

CONCLUSION

CPA may be not a so rare pulmonary malposition anomaly

as we thought before. Although it had no meaningful hemodynamics, its clinical significance lies in differential diagnosis of vascular rings and reminding clinician to do other assessments. The diagnosis of the CPA is also significant. For the consideration of children of avoiding radioactive contact, MSCT is not recommended for CPA. Echocardiography can diagnose CPA accurately as an economic, convenient, safe, non-radioactive, image modality.

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