

Retrospective Echocardiographical Analysis of Unilateral Absence of Pulmonary Artery

J CHEN, L ZHAO, ZL ZHENG, YJ ZHOU, L ZHANG, GP JIANG

Abstract

Objective: To investigate the value of echocardiography in diagnosing unilateral absence of pulmonary artery (UAPA). **Methods:** The pulmonary artery branches were examined on the parasternal aortic root short axis and pulmonary artery branch view by echocardiography; Colour Doppler was further used to confirm the absence of left pulmonary artery and the absence of right pulmonary artery. The results of echocardiography were compared with that of surgery, X-ray angiography, magnetic resonance (MR) and computed tomography (CT). **Results:** The diagnosis of UAPA in 13 patients was confirmed by surgery, X-ray angiography, MR and CT. Among the 13 UAPA patients, 11 were diagnosed initially by echocardiography, and 2 missed diagnosis. For 3 of the 11 UAPA patients, pulmonary arteries were displayed ambiguously on the parasternal aortic root short axis views, but clearly on the parasternal pulmonary artery branch views. The 11 patients had significant expansion of their single pulmonary artery, and the mean ratio of the expanded pulmonary artery diameter/the main pulmonary artery diameter was 0.83 ± 0.13 . Two UAPA patients were complicated with lung dysplasia. **Conclusion:** The parasternal pulmonary artery branch view is more potent in diagnosing UAPA than the parasternal aortic root short axis view. The ratio of the left/right pulmonary artery diameter and the main pulmonary artery diameter around 0.8 should raise consideration for UAPA. Lung dysplasia should be considered in UAPA children.

Key words

Children; Echocardiography; Unilateral absence of a pulmonary artery

Introduction

Congenital unilateral absence of a pulmonary artery (UAPA) is a rare anomaly,^{1,2} most frequently accompanied by cardiovascular anomalies such as tetralogy of fallot

(TOF) or septal defects.³ Due to intrapulmonary gas interference, the pulmonary artery is sometimes unclear in the parasternal aortic root short axis view by echocardiography. We found that the pulmonary artery can be observed very clearly on parasternal pulmonary artery branch view.

In this study, we investigated the value and characteristics of parasternal pulmonary artery branch view on diagnosis in UAPA. The results of echocardiography of parasternal aortic root short axis view and parasternal pulmonary artery branch view are both checked for 11 UAPA patients, and compared the results to that of operation, X-ray angiography, magnetic resonance (MR) and computed tomography (CT).

The Frist Affiliated Hospital, Zhejiang University School of Medicine, Hangzhou 310003, China

J CHEN (陳劍) MD
ZL ZHENG (鄭哲嵐) MD
YJ ZHOU (周逸蔣) MD
L ZHANG (張力) MD, PhD

Department of Pediatric Echocardiography, Children's Hospital, Zhejiang University School of Medicine, Hangzhou 310003, China

L ZHAO (趙鐳) MD
GP JIANG (蔣國平) MD

Correspondence to: Dr L ZHANG and Dr GP JIANG

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Methods

We retrospectively analysed 13 patients diagnosed as UAPA from January 2001 to June 2012 both from in-patient

and out-patient. All the echocardiographers in our study are qualified and experienced. Two-dimensional and Doppler echocardiography was performed using VIVID-7 (GE) and IE33 (PHILIPS) echocardiography. Apart from the routine scanning of cardiac and great vascular anomalies, the pulmonary artery and the left/right pulmonary artery from parasternal aortic root short axis and parasternal pulmonary artery branch view were also examined to observe pulmonary artery bifurcation and collateral vessels in all patients. The parasternal aortic root short axis shows the right ventricular outflow tract, pulmonary artery trunk, aortic valve, pulmonary valve and right and left pulmonary artery (see Figure 1). The parasternal pulmonary artery branch view clearly shows the pulmonary artery branch (see Figure 2). Echocardiography of the UAPA patients showed that a unilateral pulmonary artery branch was markedly dilated,

with absence of the other pulmonary artery branch on pulmonary artery bifurcation. The absence of left pulmonary artery (ALPA) is shown in Figure 3, and the absence of right pulmonary artery (ARPA) in Figure 4. The ALPA and ARPA were further confirmed by Colour Doppler Echocardiography. The results were compared with surgery, X-ray angiography, MR and CT results.

Results

A total of 13 children were diagnosed with UAPA from 2001 to 2012. One of the 13 cases was isolated UAPA (IUAPA). The average age of the 13 patients was 7.7 years old (arrang 17 days of age – 16 years). Eleven patients were diagnosed by echocardiography, and two missed by echocardiography.

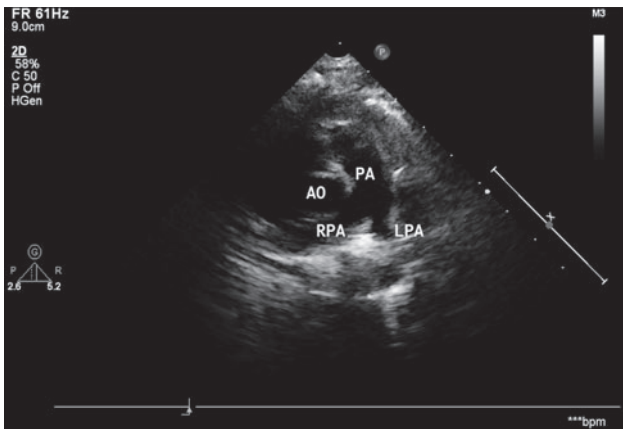


Figure 1 Aorta (AO), pulmonary artery (PA), right pulmonary artery (RPA) and left pulmonary artery (LPA) in parasternal aortic root short axis.

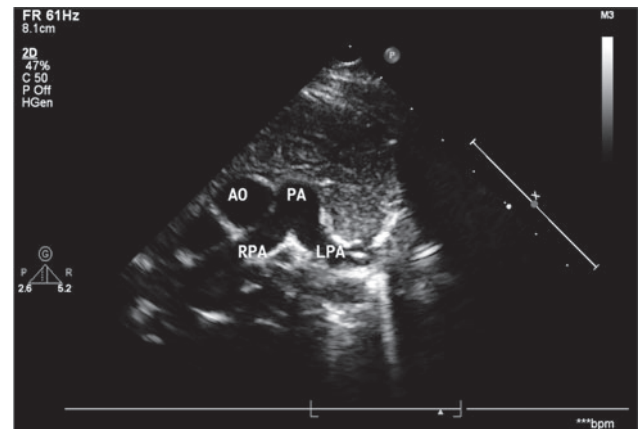


Figure 2 Pulmonary artery (PA) branch, right pulmonary artery (RPA) and left pulmonary artery (LPA) in parasternal pulmonary artery view. AO: aorta

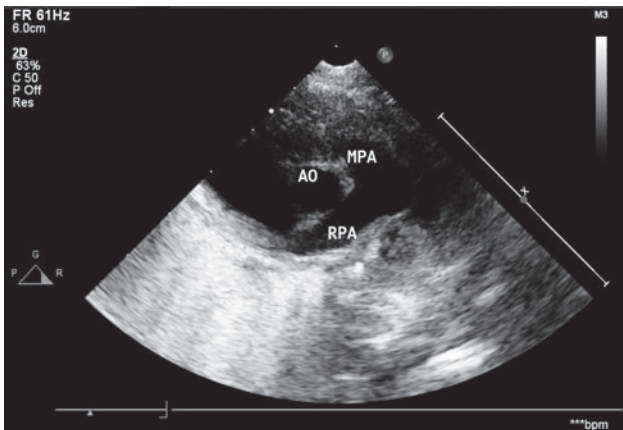


Figure 3 Absence of left pulmonary artery in parasternal pulmonary artery branch view. AO: aorta, MPA: main pulmonary artery; RPA: right pulmonary artery.

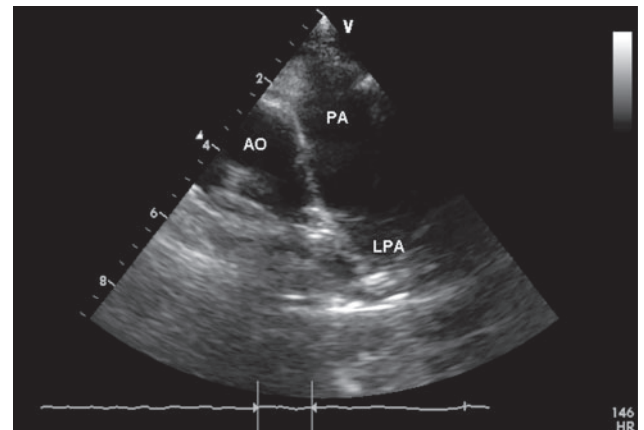


Figure 4 Absence of right pulmonary artery in parasternal pulmonary artery branch view. AO: aorta, PA: pulmonary artery; LPA: left pulmonary artery.

Among the 13 UAPA patients, 8 were confirmed by surgery, 3 by X-ray angiography, and 2 by MR and CT. The characteristics of the thirteen children with UAPA are summarised in the Table 1.

For the two patients with missed diagnosis by

echocardiography, one was due to the thick collateral vessel, and the other due to patent ductus arteriosus (PDA) mistakenly regarded as left pulmonary artery. The associated malformations of UAPA are illustrated in Table 2. In the table, two cases had lung hypoplasia in

Table 1 Characteristics of 13 patients with unilateral absence of pulmonary artery

Case (No.)	Age (Months)	Gender	Clinical Manifestation	X-ray Angiography	MR	CT	Chest X-ray	Operation
1	7	M	Cardiac murmur	TOF, ALPA, collateral vessel			Shadow of heart, lung veins increase	Yes
2	0.5	F	Hoarseness		ALPA		Shadow of heart, lung veins increase	No
3	144	M	Murmur	ASD, ALPA, PH			Shadow of heart, lung veins increase	Yes
4	156	F	Haemoptysis				Shadow of heart, lung veins increase	Yes
5	5	M	Cyanosis	TOF, ALPA, collateral vessel			Shadow of heart, lung veins increase	Yes
6	144	F	Cardiac murmur	ASD, PDA, ARPA, collateral vessel			Shadow of heart, lung veins reduce	Yes
7	59	F	Fever, cough dyspnoea	PDA, ARPA			Shadow of heart, lung veins increase	No
8	177	M	Cardiac murmur, rachioscoliosis			ARPA, right lung dysplasia	Shadow of heart, left lung veins increase, right lung dysplasia	No
9	192	F	Cardiac murmur, deformed thoracic vertebrae				Shadow of heart, lung veins increase	Yes
10	129	M	Cyanosis	TOF, ALPA, collateral vessel			Shadow of heart, left lung veins increase, right lung veins reduce	No
11	120	F	Cyanosis	PA, VSD, ASD, PDA, ALPA, collateral vessel			Shadow of heart, lung veins reduce	No
12	3	F	Fever, cough	ASD, PDA, ALPA, collateral vessel			Shadow of heart, left lung dysplasia, right lung veins reduce	Yes
13	68	M	Cardiac murmur	TOF, ALPA, collateral vessel			Shadow of heart, lung veins reduce	Yes

F: female; M: male; UPRPA: absence of right pulmonary artery; UPLPA: absence of left pulmonary artery; VSD: ventricular septal defects; ASD: atrial septal defect; PDA: patent ductus arteriosus; PA: pulmonary atresia; MR: magnetic resonance; CT: computed tomography; TOF: tetralogy of fallot

Table 2 ALPA/ARPA with cardiovascular malformations

	TOF	VSD	ASD	PDA	ASD+PDA	VSD+ASD+PDA	PA+ASD+VSD+PDA
ALPA	4		1		1	1	1
ARPA		1	1	2	1		

ALPA: absence of left pulmonary artery; ARPA: absence of right pulmonary artery; TOF: tetralogy of fallot; VSD: ventricular septal defects; ASD: atrial septal defect; PDA: patent ductus arteriosus; PA: pulmonary atresia

addition to UAPA.

All the patients showed cyanosis, shortness of breath, drop in activity tolerance, recurrent lung infection and growth delay. Physical examinations showed that twelve patients had enlarged heart and heart murmur, etc. Electrocardiogram revealed sinus tachycardia, right ventricular hypertrophy in the patients. Chest radiography results showed that enlarged heart at varying degrees; most patients had enlarged right ventricles. Chest X-ray shows increased bilateral markings in six patients, decreased bilateral markings in three patients, and decreased unilateral markings in three patients; one patient had lung hypoplasia. One case was diagnosed as left lung hypoplasia confirmed by surgery. One case was diagnosed as right lung hypoplasia confirmed by CT. Thirteen patients had pulmonary hypertension, with four patients with severe pulmonary hypertension, two of which were diagnosed as lung hypoplasia mentioned above.

Among 13 UAPA patients, eleven of them were diagnosed initially by echocardiography, and two were missed. For three of the eleven UAPA patients, pulmonary arteries were displayed unclearly on the parasternal aortic root short axis view. However, the pulmonary artery branch in the above 3 patients could be displayed clearly on the parasternal pulmonary artery branch view.

Single pulmonary arteries in all patients were significantly expanded. For all patients, the main pulmonary aortic diameter is 1.61 ± 0.68 cm. In ARPA patients, the right pulmonary artery diameter is 1.29 ± 0.72 cm, while in ALPA patients, the left pulmonary artery diameter is 1.54 ± 0.43 cm. The dimensions of the main and branch pulmonary arteries were normalised to body surface area and expressed as z score based on published normograms.⁴ The z score of main pulmonary artery dimension is 0.23 ± 1.72 , for the left pulmonary artery is 3.07 ± 1.01 , and that for the right pulmonary artery is 2.02 ± 1.91 . In total, the ratio of the expanded left/right pulmonary artery diameter/the main pulmonary artery diameter was 0.83 ± 0.13 .

Among the 13 UAPA patients, 8 were confirmed by surgery, 3 by X-ray angiography and 2 by MR and CT. Five patients refused to continuing therapy.

Discussion

Unilateral absence of pulmonary artery was reported first by Fraenzel in 1868.⁵ It accounted for about 0.1% of congenital heart diseases. UAPA may occur alone or be complicated by other cardiovascular malformations. Complex cardiovascular malformation always accompanies

with ALPA.⁶ In accordance with literature, 5 patients in this study had absence of left pulmonary arteries combined with complex cardiovascular malformations.

The 2D echocardiography is widely used in clinic. It is simple and non-invasive. Some studies showed that the diagnostic rate of echocardiography for absence of left pulmonary artery was low.⁷ It is due to several factors, including inaccessibility to pulmonary arteries, and limited ultrasonic windows for viewing pulmonary artery branch. Among the 13 UAPA patients, 11 were diagnosed initially by echocardiography. The diagnostic accuracy is 84.6%.

The parasternal aortic root short axis view is a routine view to display main pulmonary artery and pulmonary artery branches. However, it is difficult to display clearly the main pulmonary artery and pulmonary artery branch because the pulmonary gas interferes with the pulmonary arteries in some cases such as TOF, pulmonary Atresia (PA), etc. Since the relatively large thymus in children can push backward the lung tissue in front of the pulmonary artery, the pulmonary artery branch view may display the pulmonary artery and its left/right branches better than the parasternal aortic root short axis view. In our study, the image of the IUAPA patient can be displayed clearly both with parasternal aortic root short axis and parasternal pulmonary artery branch view, probably due to lack of complex anomaly and less interference with lung gas. In this study, pulmonary artery bifurcation and left/right pulmonary artery in three cases could not be observed clearly. However, the pulmonary artery bifurcation and pulmonary arteries could be displayed clearly on the parasternal pulmonary artery branch views, and they were definitely diagnosed as UAPA. The parasternal pulmonary artery branch view display the pulmonary artery bifurcation and pulmonary arteries more clearly, and thus the diagnosis of UAPA can be made more precisely.

Under normal conditions, the blood in pulmonary artery inflow to the left or right pulmonary artery equally, and diameters of left or right pulmonary artery root should be similar. In UAPA patients in our study, blood in main pulmonary artery all inflow to the single pulmonary artery, resulting in pulmonary artery expansion. In this study, for all the 11 UAPA patients diagnosed by echocardiography, the contralateral pulmonary arteries expanded apparently, with 7 patients developing pulmonary hypertension. The ratio of the expanded left/right pulmonary artery diameter/the main pulmonary artery diameter was 0.83 ± 0.13 . Thus we suggest, when the ratio of the expanded left/right pulmonary artery diameter/the main pulmonary artery diameter is around 0.8, it should be regarded as a guidance of an index that would raise suspicion of diagnosis for

UAPA.

In UAPA patients, the blood in main pulmonary artery cannot flow into the pulmonary tissues, resulting in pulmonary tissue ischaemia and hypoxia, and causing collateral vessels growth.⁸ In the lungs of UAPA patients, the feeding arteries are bronchial artery and aberrant arteries originating from the ascending or descending aorta. Moreover, it can also originate from innominate artery, intercostal artery, internal mammary artery, etc. Collateral vessels are often found at the pulmonary branch. In our study, a large collateral vessel in one child was mistakenly regarded as left pulmonary artery (Figure 5). This missed diagnosis was made because the pulmonary artery branch of TOF was not easily observed by echocardiography and the collateral vessel had been incorrectly taken as left pulmonary artery by two-dimensional and Doppler image. Using of Doppler and colour Doppler carefully and properly can improve the rate of diagnosis for TOF patients with ALPA.⁹

The anatomy and volume of lung in UAPA patients is basically normal when born. If vascular reconstruction operation can be undertaken early after birth,^{10,11} it is likely that the pathological side of the pulmonary tissue develop normally. If surgery is not taken timely to establish rich collateral circulation for blood supplying, then the pathological side of pulmonary tissue may be hypoplastic.^{8,9} In this article, 2 cases of UAPA were diagnosed with pulmonary hypoplasia by X-ray angiography and CT, and both cases complicated by severe pulmonary hypertension. This shows that attention should be paid to the hypoplasia of the pathological side of the lung when diagnosing UAPA by echocardiography. Meanwhile, if one side of lung tissue was found hypoplastic in clinical practice,

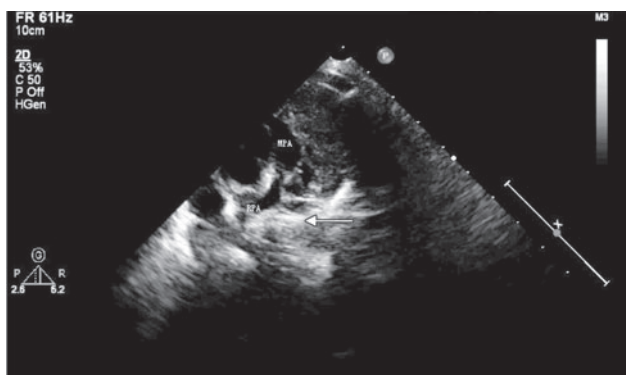


Figure 5 Absence of left pulmonary artery with tetralogy of fallot, the collateral vessel denoted by arrow had been incorrectly taken as left pulmonary artery. MPA: main pulmonary artery; RPA: right pulmonary artery.

echocardiography should be the first-choice modality to make the diagnosis. It is required to examine pathological side of pulmonary artery and UAPA by echocardiography.

Conclusion

The parasternal pulmonary artery branch view is more potent in diagnosing UAPA than the parasternal aortic root short axis view. The ratio of the left/right pulmonary artery diameter and the main pulmonary artery diameter around 0.8 should raise consideration for UAPA. Lung dysplasia should be considered in UAPA children.

Declaration of Interest

We declare that we have no conflict of interests.

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