

## Original Article

# Clinical analysis and follow up study of pulmonary artery sling in childhood and risk factors associated with mortality in nonsurgical patients

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**Abstract:** Aim: We aimed to evaluate clinical characteristics and therapeutic strategies of pulmonary artery sling (PAS) in pediatric patients and explore its risk factors for mortality in non-surgically treated children. Methods: A retrospective review of 74 patients diagnosed with PAS followed up at Guangzhou Women and Children's Medical Center from January 2011 to January 2017. For each patient, data of demographics as well as clinical presentations, imaging characteristics, family medical records, laboratory observations, treatment protocols, and outcomes were reviewed and analyzed. Results: The study group consisted of 43 boys and 31 girls (male/female ratio 1.4:1.0). The age at onset ranged from birth to 48 months (mean 3.92 months). Age at diagnosis ranged from 1 day after birth to 84 months (mean 9.23 months). Of the 74 patients, 46 underwent surgery in our hospital. The median age of surgical patients was 5.5 months (range 3-13.25 months). Regarding surgical outcome, 44 PAS children survived and 2 died, postoperatively. In the non-surgical group, 13 children died in the hospital or shortly after abandoning treatment post-discharge. Deceased patients in the non-surgical group were more likely to be younger at onset than surviving patients ( $P < 0.05$  for comparisons). Conclusions: Surgical procedures are an important and effective means of relieving left pulmonary artery compression and potentially achieving a better curative effect. Only early onset significantly influenced mortality in non-surgical patients with PAS. Patient age at the time of surgery needs to be further explored.

**Keywords:** Pulmonary artery sling, clinical characteristics, risk factors, prognosis

## Introduction

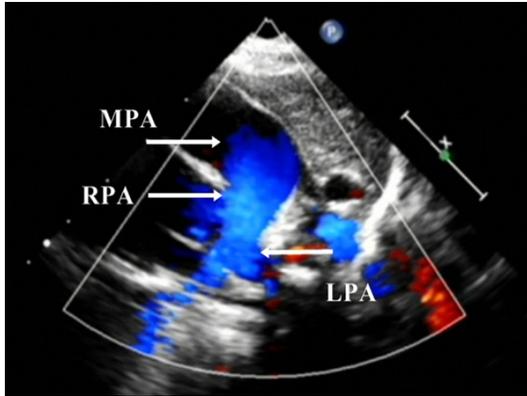
Pulmonary artery sling (PAS) is a rare congenital anomaly in which the left pulmonary artery originates from the right pulmonary artery and courses over the right main bronchus and then posteriorly between the trachea and esophagus to reach the left lung, forming a partial sling around the trachea [1]. Prevalence of PAS is reported as 1 in 17,000 school-aged children and its incidence as 0.14% among hospitalized patients with congenital heart disease [2, 3]. Clinical findings that have emerged in association with the left pulmonary artery vary according to the degree of pressure it causes on the respiratory tract and/or esophagus. It is emphasized that early identification and timely surgical treatment is the key to survival since some children have life-threatening

severe dyspnea in the neonatal period and in infancy. However, given the current state of improvement in surgical procedures and knowledge of the disease, timing of surgery and how to deal with airway stenosis remain controversial [4, 5]. Many clinicians pay more attention to risk factors for death after surgery but no studies to date have evaluated risk factors for mortality in non-surgical children with PAS. The main purpose of this survey is to present clinical and imaging characteristics and explore risk factors for mortality in children with PAS that do not undergo surgery.

## Materials and methods

### Clinical data

This study included 74 patients diagnosed with PAS that were followed up at Guangzhou



**Figure 1.** A 1-month-old girl with pulmonary artery sling (PAS), Echocardiography showed that there was no left pulmonary artery (LPA) echo at the conventional pulmonary artery bifurcation and an abnormal vessel was seen to open to the right pulmonary artery (RPA) and to the left.

Women and Children's Medical Center from January 2011 to January 2017. Diagnosis of PAS was based on routine echocardiography and computed tomography (CT). Diagnosis of tracheal stenosis was confirmed by chest CT scans and three-dimensional reconstruction and/or bronchoscopy. For each patient, data of demographics as well as clinical presentations, imaging characteristics, family medical records, laboratory observations, treatment protocols, and outcomes were reviewed and analyzed.

The study was conducted in accordance with the declaration of Helsinki and with approval from the Ethics Committee of Guangzhou Women and Children's Medical Center, Guangzhou Medical University. Written informed consent was obtained from the parents or guardians of all participants.

### Statistical analysis

All statistical analyses were performed using Statistical Package for the Social Sciences (SPSS) v.17.0. Data are described as frequencies and mean with ranges. Factors associated with mortality were compared using Fisher's exact test for a small population and Wilcoxon paired rank sum test was used to compare age at onset and age at diagnosis among non-surgical patients who died or survived. A value of  $p < 0.05$  was considered statistically significant.

## Results

### Characteristics of patients

The study group consisted of 43 boys and 31 girls (male/female ratio 1.4:1.0). Age at onset ranged from birth to 48 months (mean 3.92 months). The onset age was earlier than 1 month in 24 patients (32.43%) and before 12 months in 69 (93.24%). Age at diagnosis ranged from 1 day after birth to 84 months (mean 9.23 months) except for one case of suspected PAS during pregnancy (32 weeks), while undergoing fetal echocardiography.

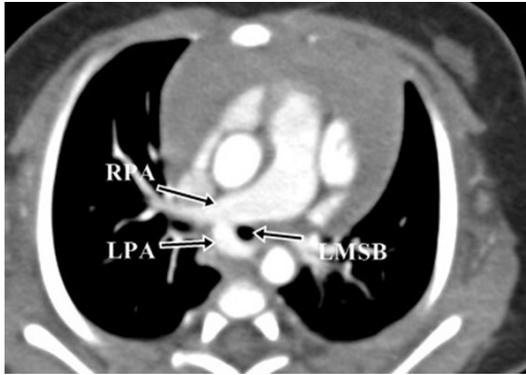
Most of the patients (57 of 74) were born at full term, the remainder had been prematurely delivered (<37 weeks). Seven of the 57 full-term infants were considered small for gestational age (weight <2.5 kg).

By the time of diagnosis, four patients were asymptomatic. The respiratory system was the most frequently involved with clinical manifestations and severity among patients varied. The majority of patients exhibited persistent or intermittent wheezing (41/74, 55.41%) and cough (39/74, 52.70%). Other common clinical findings were shortness of breath (25/74, 33.78%), stridor (15/74, 20.27%), cyanosis (9/74, 12.16%), recurrent respiratory tract infection (9/74, 12.16%), and feeding difficulty (8/74, 10.81%). Four children had a history of mechanical ventilation before diagnosis, of which two were born with acute respiratory distress syndrome.

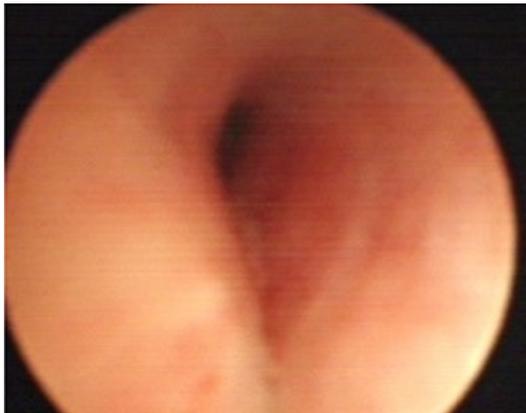
### Imaging findings at diagnosis

All children underwent echocardiography which revealed 59 cases of PAS (**Figure 1**) and 10 cases of suspected PAS. In the 5 remaining cases, PAS was later confirmed by CT. The initial diagnostic rate for echocardiography was 79.7% (59/74). Sixty-seven children underwent CT, all of whom were diagnosed with PAS (**Figure 2**) (diagnostic rate 100%). Fifty-three children underwent bronchoscopy, whereby different degrees of tracheal stenosis were found in 49 cases (**Figure 3**). Most patients had compound malformations associated with respiratory malformations (68/74, 91.89%), cardiovascular malformations (48/74, 64.86%), digestive system malformations (6/74, 8.11%), and other systemic malforma-

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**Figure 2.** A 1-month-old girl with pulmonary artery sling (PAS), Enhanced CT showed left pulmonary artery (LPA) arising from the distal right pulmonary artery (RPA) and passing behind the left main stem bronchus (LMSB) causing the bronchial compression.



**Figure 3.** Bronchoscopy showed the stenosis of the left main stem bronchus (LMSB) because of external oppression.

tions (9/74, 12.16%) revealed by echocardiography, CT, and/or bronchoscopy. Congenital abnormalities associated with PAS are shown in **Table 1**.

### *Treatment and outcome of the patients*

Forty-six of 74 patients received surgical management. Their age at the time of surgery ranged from 23 days to 84 months (mean 12.74 months). The remainder (n=28) did not undergo surgery. PAS re-implantation only was performed in 14 children while 32 patients received PAS re-implantation along with other diorthoses including cardiovascular surgery (n=28), tracheal surgery (n=4) (end-to-end anastomosis in 3 patients and sliding trach-

eoplasty in 1 patient), and isolated pneumonectomy (n=2). Postoperative time in the intensive care unit ranged from 1 to 34 days (mean 6.96 days).

Follow up data were based on outpatient or telephone information over a time span of 2-75 months (mean 28.3 months). Notably, 5 out of 28 non-surgical patients were not available for follow up investigation. Regarding surgical outcome, 44 PAS children who underwent surgery had a favorable prognosis during their follow up visit and 2 patients died postoperatively. The first death occurred 2 months postoperatively in a child that initially underwent left pulmonary artery re-implantation only, for whom tracheal stenting was necessary because of severe tracheal stenosis. The patient died of tracheal obstruction due to aggravated granulation tissue hyperplasia. The second patient died 1 month post-operatively after undergoing left pulmonary artery re-implantation and end-to-end anastomosis simultaneously. Death was due to respiratory failure caused by recurring anastomotic fistula. The majority of patients with PAS generally had continuous wheezing and other respiratory symptoms for 6 to 19 months after their operation.

Thirteen children in the non-surgical group, all younger than six months, died in the hospital or shortly after abandoning treatment post discharge from hospital. The main cause of death among these patients was severe respiratory failure. Nine patients experienced exacerbated pulmonary conditions induced by infection during follow up and required readmission to the hospital, except for one patient who is currently asymptomatic.

Analysis of the risk factors for death among non-surgical patients is shown in **Table 2**. Deceased patients were more likely to have had onset at a younger age than the surviving patients ( $P<0.05$  for comparisons).

### **Discussion**

PAS is a rare type of vascular ring that causes compression of the trachea and/or esophagus [1, 5]. External compression and intrinsic stenosis of the trachea leads to respiratory symptoms. In our series of 74 Chinese cases, the incidence rate was higher in males than in females, consistent with a previous report [6].

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**Table 1.** Congenital abnormalities associated with PAS

System	Congenital abnormal disease	No. of patients (%)
The respiratory system	Tracheobronchial stenosis	62 (83.78%)
	Bridging bronchus ( <b>Figure 4</b> )	11 (14.86%)
	Tracheal malacia	7 (9.46%)
	Pulmonary hypoplasia	5 (6.76%)
	Right upper lobe tracheal bronchus	5 (6.76%)
	Tracheal diverticulum	3 (4.05%)
	Pulmonary sequestration ( <b>Figure 5</b> )	2 (2.70%)
	Horseshoe lung ( <b>Figure 6</b> )	1 (1.35%)
The cardiovascular system	Atrial septal defect	25 (33.78%)
	Patent ductus arteriosus	21 (28.37%)
	Persistent left superior vena cava	19 (25.68%)
	Ventricular septal defect	10 (13.51%)
	Left vertebral artery origin alone	2 (2.7%)
	Mitral valve regurgitation	1 (1.35%)
	Left pulmonary artery stenosis	2 (2.7%)
	Right pulmonary artery hypoplasia	1 (1.35%)
The digestive system	Moderate pulmonary hypertension	1 (1.35%)
	Congenital anal atresia	4 (5.41%)
	Hirschsprung's disease	1 (1.35%)
Other systems	Brochopulmonary foregut malformation	1 (1.35%)
	Skeletal deformity	6 (8.11%)
	Hypospadias	2 (2.7%)
	Right ankylotia	1 (1.35%)

Most patients with PAS in our study tended to present symptoms earlier in life. About one-third of the patients had symptoms during the neonatal period while 69 (93.24%) were symptomatic in the first year of life. Moreover, respiratory symptoms were more prominent. Clinically, patients presented with wheezing, cough, progressive dyspnea, and recurrent respiratory infections although severe symptoms such as respiratory failure can occur in some cases. These findings are also consistent with those of previous reports [6-8]. In fact, children with PAS are often overlooked or remain unnoticed because of its nonspecific clinical manifestations. In fact, four of our patients were asymptomatic at the time of diagnosis and this form may be much more difficult to recognize.

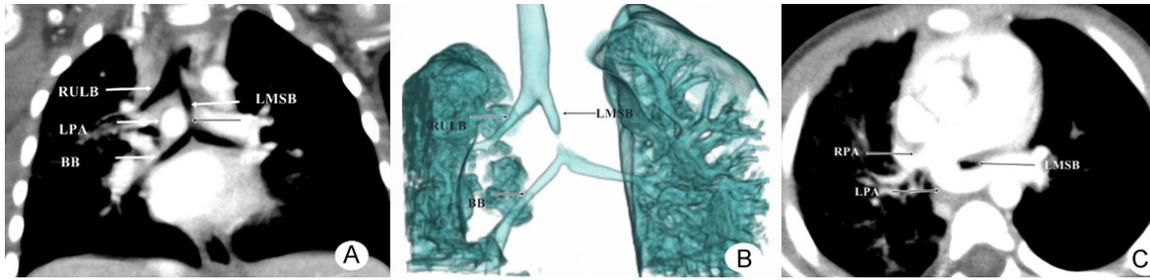
In our present study, most (91.89%) of the patients had combined respiratory and cardiovascular malformations (64.86%). The main abnormalities of the respiratory system reported here were tracheobronchial stenosis, brid-

ging bronchus, and tracheomalacia. Main abnormalities of the cardiovascular system in our patients were atrial septal defect, patent ductus arteriosus, and persistent left superior vena cava, consistent with previous literature [5, 9]. PAS commonly has varying degrees of tracheobronchial stenosis because of tracheal compression by the aberrant left pulmonary artery and/or tracheobronchial anomaly (such as abnormal cartilage rings and absent tracheal pars membranacea). Some reports have found bridging bronchus to be associated with cardiac or vascular anomalies, especially PAS [10, 11]. The embryological basis for this anomaly is not clear. Pulmonary vasculature may play some role in the

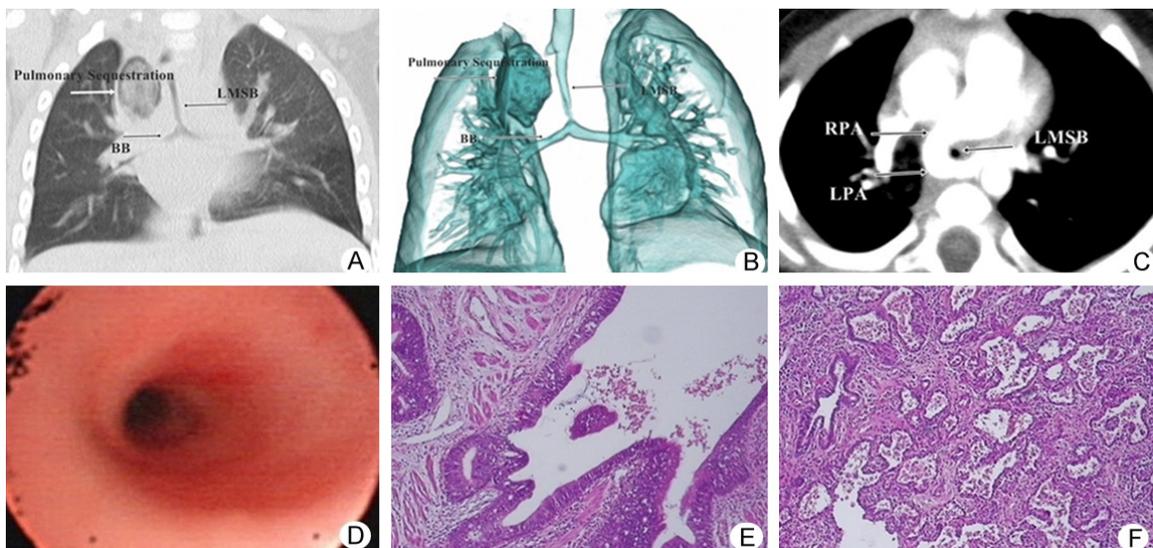
development of a bridging bronchus. An abnormal junction of the embryonic left pulmonary artery with the pulmonary plexus occurs during the fifth gestational week, giving rise to PAS [12]. Patent ductus arteriosus and persistent left superior vena cava formation has been explained by the "space availability" theory [9]. However, the mechanism behind formation of other malformations is still unclear. There is evidence of genetically related factors, such as was reported in a case of PAS in identical twins [13] and in patients with trisomies 18 and 21 [14, 15]. Thus, given that the cause of PAS and concomitant malformations is multifactorial, patients with congenital cardiovascular anomalies should be evaluated for tracheobronchial anomalies.

Diagnosis of PAS mainly depends upon imaging findings. Echocardiography, the most important auxiliary examination, can find not only PAS but also other cardiac malformations via noninvasive and nonionizing radiation [16, 17]. Some reports have pointed out that echocar-

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**Figure 4.** A 1-year-old girl with pulmonary artery sling (PAS), bronchial bridge (BB), and left main stem bronchus (LMSB) stenosis, (A) Computer tomography (CT) showed tracheal was divided into the right upper lobe bronchus (RULB) and LMSB in the third thoracic level. The right middle lobe and lower lobe bronchus originate in the LMSB at about the sixth thoracic level and across the mediastinum which was called bronchial bridge (BB). The left pulmonary artery encircled the left main stem bronchus and lead to the stenosis (black arrows). (B) CT airway reconstruction showed tracheal was divided into the RULB and LMSB, the right middle lobe, and lower lobe bronchus originate in the LMSB and the LMSB stenosis. (C) Enhanced CT showed left pulmonary artery (LPA) arising from the distal right pulmonary artery (RPA) and passing behind the left main stem bronchus (LMSB) causing the bronchial compression.

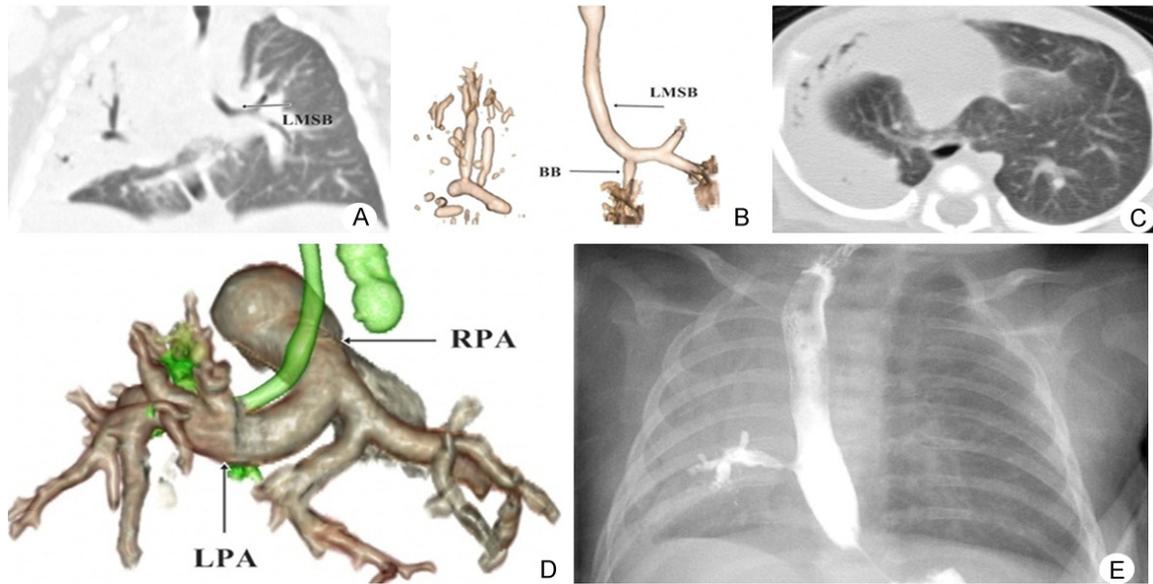


**Figure 5.** A ten-month-old boy with pulmonary artery sling (PAS), bronchial bridge (BB), and left main stem bronchus (LMSB) stenosis and pulmonary sequestration. A. Computerized tomography showed the right upper lobe bronchus was missed. The right middle lobe and lower lobe bronchus originated in the LMSB at the sixth thoracic level and across the mediastinum which was called (BB). An oval abnormal density shadow with the size of about 3\*2\*2 centimeter was seen in the right upper lung. B. CT airway reconstruction showed the right upper lobe bronchus was missed. The right middle lobe and lower lobe bronchus originated in the LMSB and the middle of the LMSB was stenosis. C. Enhanced CT showed left pulmonary artery (LPA) arising from the distal right pulmonary artery (RPA) and passing behind the LMSB causing the bronchial compression. D. Bronchoscopy showed the stenosis of the LMSB. E, F. Pathology (hematoxylin-eosin staining, 10×4 higher magnification) showed the mass was lung tissue and tracheal branch can be seen. The different sizes of small cyst with ciliated columnar epithelium and mucus pus can be seen in the lung tissue. The interstitial fibrous tissue was hyperplasia with a large number of lymphocytes, plasma cells.

diography lacks the ability to image the airway well, vessels close to the lung being particularly poorly visualized. Therefore, this method should not be used to rule out the presence of PAS [18]. Pu et al. found that diagnosis may be missed by echocardiography, especially when the anatomy is considerably distorted, as

in right lung agenesis [19]. The initial diagnostic rate for echocardiography is 79.73% in our study, similar to that previously identified in 81%-88% of cases [7, 8]. Nevertheless, echocardiography is irreplaceable in the assessment of intracardiac malformations. CT and three-dimensional reconstruction can visually

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**Figure 6.** A one-month-old girl with pulmonary artery sling (PAS), bronchial bridge (BB) horseshoe lung, and bronchopulmonary foregut malformation. A. Computerized tomography showed the right main stem bronchus was not seen in the normal tracheal carina, the distal trachea formed the left main stem bronchus (LMSB). The right middle lower lobe bronchus originated in the LMSB at about sixth thoracic level and across the mediastinum which was called BB. B. CT airway reconstruction showed the right main stem bronchus was missed and the distal trachea formed the LMSB. The right middle lower lobe bronchus originated in the LMSB. C. The bilateral lower lobes were fused in front of the spine formed the horseshoe lung. D. CT vascular reconstruction showed the left pulmonary artery (LPA) arising from the distal right pulmonary artery (RPA). E. The esophageal dynamic radiography showed the tracheo-esophageal fistula where a fistula connected the right side of the esophagus and the right lung. So, bronchopulmonary foregut malformation was diagnosed in this patient.

**Table 2.** Risk factors for death in non-surgical patients

Factor	The survival group (n=10)	The death group (n=13)	<i>p</i>
Gender (boy: girl)	5/5	9/4	0.417
Pre-term birth (Y/N)	2/8	2/11	1.00
Onset age (month)	5.00 (0.18-8.25)	0.36 (0.03-1.00)	0.025
Diagnosis age (month)	6.00 (1.52-11.5)	2.00 (1.00-4.00)	0.098
Small for gestational age (Y/N)	2/8	1/12	0.56
Malnutrition (Y/N)	1/9	2/11	1.00
Merge cardiovascular malformation (Y/N)	6/4	10/3	0.65
Merge respiratory malformation (Y/N)	10/0	11/2	0.486
Merge other systems' malformation (Y/N)	2/8	5/8	0.405
Merge more than two systems' malformation (Y/N)	6/4	10/3	0.65

show the course of abnormal blood vessels, degree of tracheal compression, and range of stenosis. Therefore, this is the best method for clinical diagnosis of PAS [18, 20]. Although CT has many advantages in diagnosing PAS, the fact that it is unable to show intracardiac abnormalities is a drawback. Bronchoscopy is the gold standard for diagnosis of tracheobronchomalacia and is very useful for dynamic

airway evaluation and assessment of the location and severity of airway stenosis [21]. However, it may be difficult in a child with severe respiratory distress which can increase edema, resulting in even further narrowing of an already compromised respiratory tract. Therefore, diagnostic work-up for PAS should include CT with multiplanar reconstructions, echocardiography to exclude cardiac and some

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vascular abnormalities, and bronchoscopy for definitive diagnosis and evaluation of tracheal stenosis and tracheobronchomalacia in the stable patient.

Some scholars have considered that PAS should not be managed medically because of life-threatening severe dyspnea in the neonatal period and infancy [22, 23] and the possibility that a delayed operation can lead to severe tracheomalacia [24]. Yu et al. indicated that early surgery can improve pulmonary function performance [2]. However, a previous study found that some patients with PAS can survive until adulthood in the absence of surgical treatment [25]. Risk and mortality were both high in relation to early surgical treatment. Some studies have already reported that mortality was highest for patients operated on in infancy because of serious complications [26, 27]. So, although we agree that surgical repair of PAS is recommended, age at the time of surgery needs to be further explored for asymptomatic patients. Current surgical management often involves concomitant repair of coexisting cardiac lesions and tracheal stenosis. The natural history of PAS is poor, with death resulting from airway obstruction. Although with the advent of the slide tracheoplasty technique mortality can undoubtedly be reduced, the mortality of PAS is determined by the need for tracheal surgery [5, 26, 28]. Some authors have suggested that the associated tracheal stenosis is best handled simultaneously [5, 7]. Others have reported that the majority of PAS with tracheal stenosis in children, who did not undergo tracheal surgery, showed no clinically significant airway problems and concomitant repair of coexisting cardiac anomalies was performed with no added mortality at their last follow up visit [4]. In the present study, 28 of our patients underwent repair of associated cardiac anomalies and only 4 had tracheal surgery. The two deaths were caused by tracheal obstruction due to aggravated granulation tissue hyperplasia and respiratory failure due to repeated anastomotic fistula after tracheal end-to-end anastomosis. Thereby, it appears that tracheal surgery is an important factor in high mortality among children with PAS. Many authors have recommended that tracheal intervention is indicated in PAS children. In Yong's study, selection of patients requiring tracheal surgery was based on

difficulty with ventilation and/or inability to wean off mechanical ventilation [26]. Another report found that a trachea diameter <3 mm was associated with use of tracheoplasty [28], while Hong et al. suggested that the diameter/length ratio was a more reliable indicator than diameter for evaluation of tracheal intervention in the first operation for PAS [29]. The children who survived without tracheoplasty in our study had varying degrees of improvement in respiratory symptoms over the follow up period, indicating that tracheal surgery is not necessary in the majority of PAS children. Indications for tracheal surgery, therefore, require further exploration in patients with PAS.

In our study, 13 of 23 non-surgical PAS patients died during the follow up period with a mortality rate as high as 56.52%. The age of all children who died was less than 6 months. Previous reports have stated that, in symptomatic infants with PAS, mortality approaches 90% without surgical intervention [6, 30]. This overall mortality is much higher than in our study, presumably due to the historical lack of knowledge about PAS and the fact that our case series was followed up for a relatively short period. However, it is regrettable that there have been no reports on the mortality of non-surgical patients with PAS in recent years. In the present study, numerous patients in the non-surgical group with severe tracheal stenosis and multiple malformations such as congenital anal atresia required complex surgery but their parents abandoned treatment because of unpredictable surgical complications and high medical costs. These patients eventually died of severe respiratory failure. In addition, the deceased non-surgical patients were more likely to be younger at onset than those who survived ( $P < 0.05$  for all comparisons). These results imply that younger onset age might potentially be a risk factor for death in children with PAS that do not undergo surgery.

### Conclusions

When a clinician is presented with a neonate or infant who has wheezing, stridor, and shortness of breath with poor efficacy to regular treatment, there should be high alertness for PAS. Definitive diagnosis of PAS requires the combination of different imaging modalities.

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Only early age onset significantly influences mortality in non-surgical patients with PAS. Surgical procedures are an important and effective means of relieving left pulmonary artery compression and potentially achieving a better curative effect. The majority of PAS children with tracheal stenosis do not require tracheal surgery and age at the time of surgery needs to be further explored.

### Acknowledgements

This research was not supported by any specific grant from funding agencies. This study was conducted in accordance with the Declaration of Helsinki. This study was conducted with approval from the Ethics Committee of Guangzhou Women and Children's Medical Center, Guangzhou Medical University.

Informed consent was obtained from all of the individual participants included in the study.

### Disclosure of conflict of interest

None.

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