Congenital tracheal stenosis with pulmonary artery sling: 3 case reports and literature review

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Pulmonary artery sling is a rare vascular anomaly causing respiratory distress in young infants. It is frequently associated with the anomalies of tracheobronchial tree which may lead to fatal outcome. We reported 3 cases of long-segment tracheal stenosis with left pulmonary artery sling (LPAS). All the 3 cases had presenting symptoms in the first year of life, 2 cases with noisy breathing and 1 case with severe upper airway obstruction and respiratory failure. The investigations performed for diagnosing LPAS and associated tracheal stenosis included plain chest radiographs, barium esophagogram, fiberoptic laryngobronchoscopy, computerized tomography of the chest and pulmonary angiography. One patient who had severe upper airway obstruction and respiratory failure underwent tracheal resection with end-to-end anastomosis and reimplantation of LPAS with a satisfactory outcome. It should be emphasized that complete evaluation of possible associated anomalies is essential for appropriate management in infants with persistent noisy breathing caused by vascular abnormalities.

Keywords: Tracheal stenosis, Pulmonary artery sling.

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ความผิดปกติของเส้นเลือดบอดเป็นความผิดปกติเดิมที่เกิดก่อนรุ่นที่นำไปให้เกิดอาการมีปัญหาทางระบบหายใจในเด็กเล็กที่พบได้ไม่บ่อย แต่มีพบร่วมกับความผิดปกติของหลอดลม ซึ่งอาจทำให้มีอาการจุดเด่นทางเดินหายใจรุนแรงถึงเสียชีวิตได้ ได้รายงานผู้ป่วย 3 รายที่ได้รับการรักษาอย่างเป็นหลักตามแต่ก่อนรวมกับความผิดปกติของเส้นเลือดบอดช่วย ผู้ป่วยทุกรายแสดงอาการในช่วงปีแรก หรือก่อนปีที่ 2 รายมีอาการเสี่ยงทางใจค่อนข้างเฉพาะกิจ ราย 1 รายมีอาการทางเดินหายใจดุดตัน รวมกับภาวะหายใจสั้น เหลาะ การตรวจสอบข้อต่อเป็น ผลิตภัณฑ์, computerized tomography ของทางเดินและ pulmonary angiography เป็นสิ่งจำเป็นในการรักษาความผิดปกติเดิมที่เกิดจากภาวะหลอดลมที่ดีแบบเก็บเส้นเลือดที่ผิดปกติ ในรายงานนี้มีผู้ป่วย 1 ราย ที่มีอาการทางเดินหายใจสั้นบ่อยกับรุนแรงมีการหายใจสั้น เหลาะ ซึ่งอาการดังกล่าวหลังได้รับการรักษาคืนคืนให้กลับเป็นหลอดสมบูรณ์และกลับไปดี แต่ไม่ได้เห็นหลอดบอดช้างซ้าย และไม่มีภาวะหลอดข้อมหลังใต้ ในการประเมินผู้ป่วยที่ดีส่งทำความผิดปกติของเส้นเลือดใหญ่มีทางไปร่วมกับอาการเสี่ยงได้ยิ่งๆ ครั้งวิจัยเพิ่มเติม เพื่อหาความผิดปกติเดิมที่เกิดกับหลอดต่างๆที่มีหลอดร่วมด้วย เพื่อจะได้พิจารณาให้การรักษาที่เหมาะสมและมีประสิทธิภาพต่อไป
Compression of the trachea and bronchi can be caused by developmental abnormalities of pulmonary vessels and major branches of the aorta. These lesions, which generally arise from failure of abnormal regression of one or more segments of the early fetal paired aortic arch system, can produce substantial distortion of the trachea and large bronchi.\(^1\)

Left pulmonary artery sling (LPAS) in which the left pulmonary artery arises from the posterior aspect of the right pulmonary artery, courses posteriorly to the right of the trachea and passes between the trachea and esophagus to reach the left hilum (Figure 1), is a rare vascular anomaly that causes respiratory distress in infants. A new classification of LPAS basing upon the level of tracheal bifurcation and the presence or absence of an eparterial right upper lobe bronchus has been proposed.\(^2\) LPAS has been considered fatal in infants with an overall mortality of 50%, even after surgical correction.\(^3\)

The LPAS is frequently associated with the anomalies of tracheobronchial tree and congenital cardiac defects. The incidence of associated tracheobronchial tree anomalies is about 40%. These anomalies include abnormal distribution of cartilage in the walls of the trachea and major bronchi, intrinsic stenosis and abnormal branching.\(^4\) The incidence of associated cardiovascular anomalies varies from 30-80%.\(^4,5\) The most important prognostic factor is associated with the anomalies of tracheobronchial tree, especially a long segment of tracheobronchial stenosis, rather than cardiac defects. Proper evaluation of the associated tracheobronchial anomaly is essential for planning of management in LPAS.\(^4-6\)

**Case reports**

**Case 1**

An 11-month-old Thai-Korean female infant was admitted to King Chulalongkorn Memorial Hospital with a 3-day history of fever, continuous and progressive noisy breathing, productive cough and respiratory distress. The noisy breathing seemed to be associated with the secretions in her throat and was not related to meal or posture. The patient later developed respiratory distress. No history of foreign body aspiration nor chest injury was obtained.

She was normally born in Korea with a birthweight of 3 kg. Significant noisy breathing without respiratory distress had been recognized since 7 months of age. She had pneumonia which required hospitalization at the ages of 7 and 10 months. Her respiratory symptoms became prominent with coryza. However, her growth and development appeared normal.

Physical examination revealed a fully conscious infant with moderate respiratory distress.

![Figure 1. Pulmonary artery sling. The left pulmonary artery arises from the right, encircling the trachea and passing between the trachea and esophagus. This produces a right-sided compression of the lower trachea and the right main bronchus.](image-url)
The respiratory rate was 42/min. No pallor, no jaundice and no cyanosis were detected. Chest auscultation revealed good air entry, inspiratory-expiratory stridor, and generalized coarse crepitation. Others findings were unremarkable. Her oxygen saturation on room air was 97%.

A chest radiograph revealed perihilar infiltrations, wide mediastinum, high level of carina, narrow tracheobronchial air column and hemivertebra of the seventh cervical spine. Fiberoptic laryngobronchoscopy demonstrated a circumferentially narrow trachea extended from mid-trachea to the carina, with an extraluminal pulsatile compression at left anterolateral aspect of the trachea. Only the 2.2 mm sized fiberoptic bronchoscope could be passed through the stenotic site to the carina. A barium esophagogram showed a focal narrowing of the mid-thoracic esophagus due to an anterior indentation. A computerized tomography of the chest demonstrated an abnormal left pulmonary artery originated from the normal right pulmonary artery. The lower half of the cervical trachea and the entire segment of the intrathoracic trachea were narrow with the internal diameter of 3 mm. The carina was found at T 6-7 level. The hemivertebra of T2 vertebral body was also noted. Pulmonary angiogram confirmed left pulmonary artery originated from right pulmonary artery and crossed anteriorly to esophagus.

The patient was treated with 10-day-course of parenteral antibiotic, supplemental oxygen, nebulized bronchodilator and chest physical therapy. She was afebrile and able to feed a few days later. Her respiratory symptoms gradually subsided in 4 weeks. The patient was rehospitalized one month later because of recurrent pneumonia. However, she was clinically improved after 2 weeks of the supportive care and then finally went back to Korea.

**Case 2**

A previously healthy 6-month-old Thai male infant was admitted to King Chulalongkorn Memorial Hospital for noisy breathing and respiratory distress. Three weeks ago, he was diagnosed as pneumonia which required hospitalization at a provincial hospital for 5 days. His noisy breathing had been noted by his parents since then. It was not related to meal or posture. He was readmitted at the same hospital for his noisy breathing for another week without any improvement. He was then referred to our hospital.

The patient was normally born with a birthweight of 3.3 kg. No postnatal respiratory problem was noted. His growth and development were normal.

Physical examination revealed an active infant with a respiratory rate of 40/min. He had suprasternal and substernal retractions. No pallor, no jaundice and no cyanosis were noted. Inspiratory-expiratory stridor and generalized coarse crepitations were detected on chest auscultation. Others were unremarkable. His oxygen saturation on room air was 98%.

A chest radiographs revealed perihilar infiltrations, wide mediastinum, and narrow tracheobronchial air column (Figure 2). A barium esophagogram revealed an anterior indentation of mid-esophagus at the level of T5-T6 (Figure 3). A fiberoptic laryngobronchoscopy demonstrated a circumferential narrowing of the mid-trachea. Only 2.2 mm sized fiberoptic bronchoscope could be passed through the stenotic site. A computerized tomography of the chest demonstrated an abnormal left pulmonary artery originated from the normal right
Figure 2A, 2B. Chest radiographs in AP (2A) and lateral view (2B) revealed perihilar infiltrations, widening of mediastinum, and poorly identified lower tracheobronchial air column (arrow).

Figure 3. Barium esophagogram revealed an extrinsic compression on anterior wall of the mid-esophagus at T5-T6 level (arrow).

pulmonary artery, coursed to the left between the trachea anteriorly and the esophagus posteriorly. The entire segment of intrathoracic trachea was narrow. The carina was found at the level of T5-6 which was lower than normal. The cervical trachea was normal (Figure 4, 5).

Figure 4. Three-dimensional computerized tomography of the chest showed the narrowing segment of the trachea extended from the level of thoracic inlet down to the low-lying tracheal bifurcation at T5-6 level (arrow A) which was lower lying than usual. The cervical trachea was normal (arrow B).
of neuromuscular blocking agent, he developed progressive hypercapnia (maximum $PCO_2$ 182 mmHg) and hypoxemia. He was then referred to King Chulalongkorn Memorial Hospital. One day prior to referring, he developed generalized tonic-clonic seizure and hypotension which were treated with anticonvulsant and inotropic drugs, respectively.

His past history was unremarkable except for a noisy breathing since birth which was diagnosed as laryngomalacia at one week of age. His noisy breathing persisted and became worse when he had acute upper respiratory tract infection, his growth and development were normal.

Physical examination revealed an intubated infant with the body temperature of 36.8°C, blood pressure 115/60 mmHg, pulse rate 130/min and respiratory rate 40/min. Despite being on 100 % oxygen via resuscitating bag, he looked cyanotic with the arterial oxygen saturation of 30 %. Inspiratory-expiratory stridor with poor air entry was heard on chest auscultation. Others were unremarkable.

A chest radiograph revealed perihilar infiltrations, wide mediastinum and narrow tracheobronchial air column. A fiberoptic laryngobronchoscopy demonstrated a circumferential narrow trachea, 2 cm in length (50 % of intrathoracic tracheal length), down to the carina. Only the 2.2 mm sized fiberoptic bronchoscope could be passed through the stenotic part (Figure 6). Both of the main bronchial openings were normal. A computerized tomography of the chest demonstrated an abnormal left pulmonary originated from right pulmonary artery and encircled the lower trachea as well as the right main bronchus. This caused narrowing of the trachea at the level of 2.5 cm. above the carina. Pulmonary angiogram confirmed abnormal

Case 3

A 7-month-old Thai male infant was hospitalized at a provincial hospital with severe croup. He developed respiratory failure and required mechanical ventilation for over a week. Despite being on a very high setting of ventilator and high dose

Figure 5. Computerized tomography of the chest with contrast revealed abnormal left pulmonary artery (arrow 1) originated from the normal right pulmonary artery, coursed to the left between the trachea (arrow 2) and esophagus.

Supplemental oxygen, nebulized bronchodilator and chest physical therapy were given. His respiratory symptoms were gradually improved and he was discharged after one week. Four months later, he was readmitted with recurrent pneumonia. His respiratory symptoms gradually improved with supportive treatment but inspiratory stridor remained. No respiratory distress was noted during follow-ups and no hospitalization was needed.
left pulmonary artery from right pulmonary artery aligned posterior to trachea caused a long segment of tracheal stenosis, and secondary pulmonary hypertension.

The patient was strictly put in hyperextended neck position. He required sedative, neuromuscular blocking agents and high setting of mechanical ventilation prior to the surgical correction. Four days later, the operation consisting of tracheal resection with end to end anastomosis, ligation of the patent ductus arteriosus and reimplantation of left pulmonary artery were performed. The postoperative period was uneventful and the mechanical ventilator could be weaned off one week later.

Discussion

LPAS commonly causes airway obstruction during the first year of life. However asymptomatic cases have been reported in older children and adults. More than 50% of the pediatric cases present with symptoms related to the compression of the adjacent structures such as tracheobronchial trees and esophagus. Patients may present with stridor or recurrent respiratory infections due to tracheobronchial trees obstruction. Some may have dysphagia and vomiting due to esophageal obstruction. Other symptoms due to associated cardiovascular anomalies are also observed. The prevalence of asymptomatic LPAS which is much more difficult to be recognized was reported to be greater than 18%. All of our 3 reported cases were diagnosed during infancy period by the symptoms and signs of upper airway obstruction. Associated tracheobronchial and cardiovascular anomalies might lead to the early diagnosis.

Wells et al. (2) classified LPAS into 2 types basing upon the level of the carina (type 1 T4-5, type 2 T5-6). Each type was further subclassified into two subtypes according to the presence (subtype A) or absence (subtype B) of the eparterial right upper lobe bronchus. This classification is useful for surgical planning. Patients with LPAS type 2 commonly have varying degrees of tracheal stenosis due to abnormal cartilaginous rings and absent tracheal pars membranacea. They also have abnormally low tracheal "bifurcation" (pseudocarina) at the level of T6 with an increased bronchial angles or "inverted T" pattern. The level of the anterior esophageal indentation caused by LPAS is lower in LPAS type 2 when compared to type 1. The incidence of LPAS type 2B is twice when compared to that of type 2A. In our report, all three cases had long segment of tracheal stenosis associated with LPAS type 2.

After the corrective surgery, the survivors usually became asymptomatic while the expired cases were found to have no blood flow through
the reimplanted left pulmonary artery. Therefore, postsurgical complication is one of the important prognostic factors.

The morbidity and mortality depended primarily on the associated airway and cardiovascular anomalies. The patients with long-segment tracheal stenosis (the narrowing segment is longer than 30% of tracheal length) due to complete cartilaginous rings (ring-sling complex) often have the stenotic part extended beyond the contact part with the LPAS. Normal tracheal growth among these patients is impossible. Moreover, the surgical outcome after vascular correction is often unsatisfactory compared to those with isolated stenosis at the point contacted with the LPAS. The associated cardiovascular lesions consisting of 30-80% of the patients were also the important prognostic factors. The lesions included persistent left SVC, atrial and ventricular septum defects, patent ductus arteriosus, tetralogy of Fallot, common ventricle and coarctation of aorta. Gastrointestinal malformations such as imperforate anus, biliary atresia and Hirschsprung's disease, were also reported. One of our reported cases had associated patent ductus arteriosus. However, none of them had associated gastrointestinal anomalies. Among the reported associated abnormalities, long segment tracheal stenosis was associated with the high morbidity and mortality.

Therefore, it is essential to look for the accompanying tracheobronchial anomalies in the patients who have LPAS.

Traditional imaging studies commonly used for diagnosing pulmonary sling include chest radiograph, barium esophagogram, pulmonary angiography and echocardiography. The suggestive findings of plain radiography include (1) unequal aeration, recurrent pneumonia or atelectasis which is related to the intensity of tracheobronchial compression, (2) mass at the right side of the lower trachea, (3) low left hilum, (4) deviation of the lower trachea and carina to the left and (5) diminished size of the left pulmonary artery branches.

A smooth, round indentation on the anterior wall of the esophagus at the level of the carina demonstrated on lateral and oblique views of barium esophagogram is usually a reliable sign for diagnosing LPAS. However, these findings on the chest roentgenogram and barium esophagogram are nonspecific and can be found in bronchogenic cyst, esophageal duplication and enlarged mediastinal nodes.

Pulmonary angiography does not adequately disclose the nature and extent of the airway anomaly or its relationship to LPAS. It may be helpful in the cases suspected for associated cardiac anomalies. Echocardiography has several limitations in defining the relationship of the vascular structures to the airway. This is due to the limitation of the technique and intrathoracic air interference. In many patients, more advanced imaging studies such as helical CT scan of the chest with contrast, electron beam tomography (EBT) or MRI of the chest are needed for the definite diagnosis.

Tracheobronchography was previously considered as an important diagnostic modality for assessing tracheobronchial anomalies. Bronchoscopy is informative in defining airway anatomy and identifying the sites of extrinsic compression. However, both of them can be hazardous in patients with severe respiratory compromise.
diagnosis can currently be established by the non-invasive investigations, such as conventional and helical CT scan of the chest, EBT and MRI of the chest.

To date, there is still no effective treatment in severe tracheal stenosis. Forceful dilation of the trachea may result in splitting of the complete cartilaginous rings but risks for tracheal perforation or restenosis. Short-segment congenital tracheal stenosis can be successfully corrected with tracheal resection and end to end anastomosis. For long-segment congenital tracheal stenosis, there are 3 techniques used for surgical corrections. The first technique is tracheal resection with end-to-end anastomosis. The reconstructed trachea has been experimentally and clinically demonstrated to have satisfactory growth.\(^{(26,27)}\) However, only 25% to 30% of the entire tracheal length can be resected and successfully end-to-end anastomosed. Too long resected segments risk for excessive tension and suture separation at the anastomotic site.\(^{(28)}\) In our third reported case, despite having a long-segment tracheal stenosis, he had a satisfactory surgical outcome with this technique. The second technique is tracheal reconstruction with costal cartilage graft\(^{(29)}\) or pericardial patch.\(^{(30)}\) These techniques require airway stenting with intubation during the early period of tracheal wound healing. Multiple debridements of the granulation tissue at the graft sites may be required. Necrosis and collapse of the grafts were reported in some patients.\(^{(31,32)}\) Reoperation was necessary in 7 of 28 patients in one report.\(^{(33)}\) The third technique is slide tracheoplasty.\(^{(34)}\) With this technique, the circumference of the trachea can be doubled, and the cross-sectional area of the tracheal lumen can be quadrupled. The stenotic segment is shortened by half.\(^{(35)}\) The reconstructed trachea contains the native cartilages and is lined with normal tracheal epithelium. Therefore, satisfactory subsequent tracheal growth was experimentally and clinically demonstrated.\(^{(36-38)}\)

**Conclusion**

Despite being a rare vascular anomaly, LPAS should be considered in infants with persistent noisy breathing which was not related to posture or meals, wide mediastinum and narrow tracheobronchial air column demonstrated on chest x-ray. The narrow airway can be due to the external compression caused by abnormal pulmonary vessels or the associated tracheobronchial anomalies. The symptoms of airway obstruction vary from moderate respiratory distress occurring only during respiratory tract infections to severe airway obstruction and respiratory failure requiring immediate surgical correction as reported in one of our patients. Therefore, early recognition and complete evaluation of the associated tracheobronchial anomalies are essential for appropriate treatment in order to reduce the morbidity and mortality among these patients.

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