Supportive pericardial suspension for surgical airway management of tracheobronchomalacia in unilateral pulmonary agenesis

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Unilateral pulmonary agenesis, a rare developmental defect of the lung, is often accompanied by tracheobronchial stenosis or malacia due to displacement, distortion, and compression of the surrounding great vessels. We present two cases of unilateral pulmonary agenesis complicated by tracheobronchial problems that were successfully managed surgically with supportive pericardial suspension. ***Ann Pediatr Surg*** **11:147–149 © 2015 Annals of Pediatric Surgery.**

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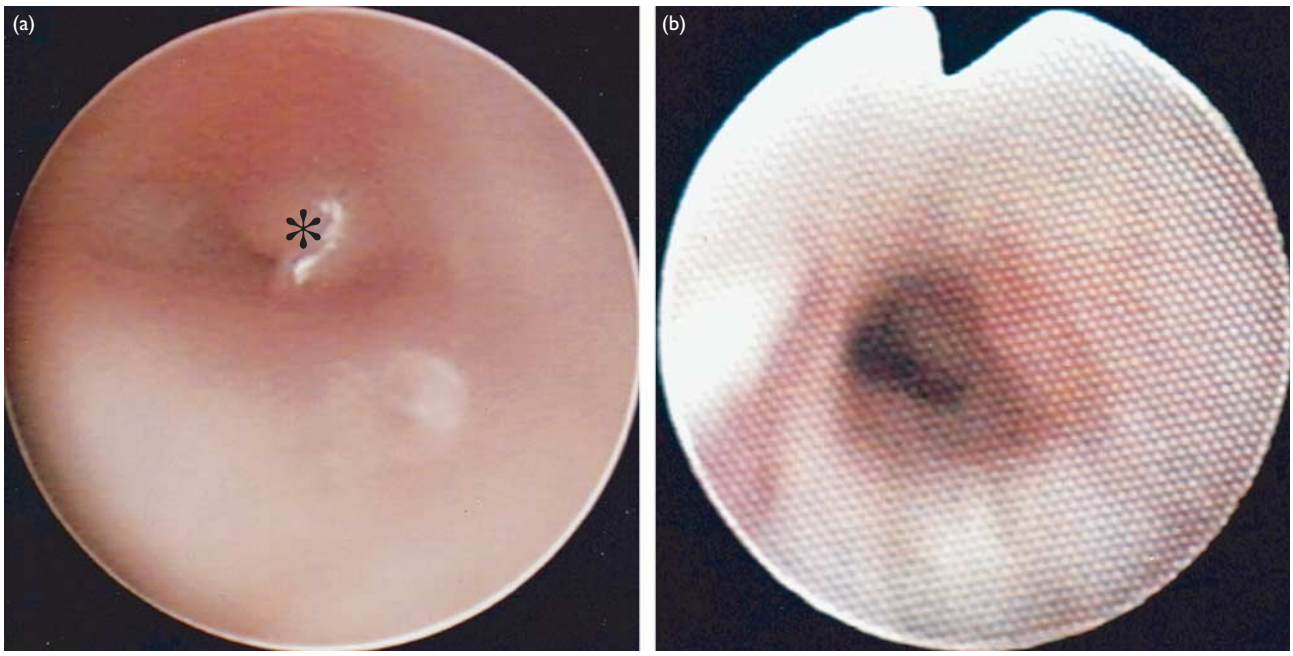
Unilateral pulmonary agenesis (UPA) is a rare developmental defect of the lung, which has been defined as a complete defect of the pulmonary parenchyma and artery with absence of the bronchus. Patients with UPA present unique anatomic features such as an ipsilateral shift and rotation of the heart and mediastinum to the empty hemithorax, which results in displacement, distortion, and compression of the great vessels and airway. Tracheobronchial stenosis and malacia often accompany UPA and are associated with high morbidity and mortality [1–3]. We report here two cases of UPA with tracheobronchial problems that were successfully managed surgically using supportive pericardial suspension.

Case 1

A male infant was born with a birth weight of 3362 g at 38 weeks of gestation. Soon after birth, he was presented with respiratory distress and required immediate intubation and mechanical ventilation. The infant was successfully extubated after improvement of persistent pulmonary hypertension at 25 days of age, but he was intubated again because of CO_2 retention with a PCO_2 exceeding 70 mmHg. Chest radiograph revealed opacity of the right hemithorax, with a shift of the mediastinum and heart to the right side. Flexible bronchoscopy showed severe malacic narrowing of the left main bronchus. Because several episodes of respiratory distress had

Figure 1



Bronchoscopic findings of case 1. (a) Preoperative rigid bronchoscopic image showing complete obstruction of the left main bronchus with granulation tissue (*). (b) Intraoperative flexible bronchoscopic image showing adequate luminal opening of the left main bronchus after arteriopexy with pericardial suspension.

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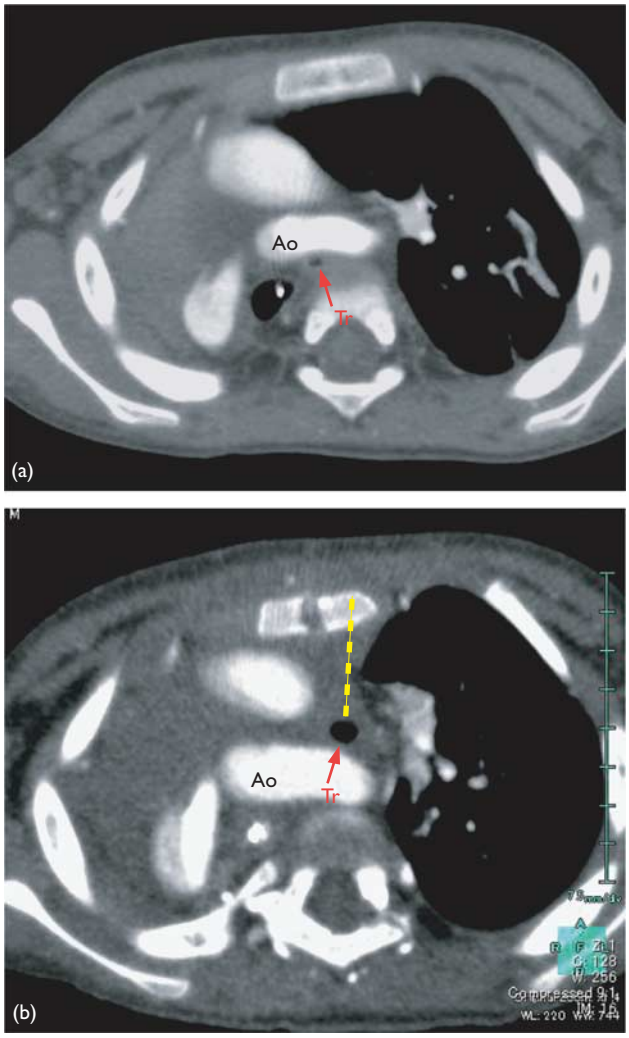
occurred, a tracheostomy was performed at 3 months of age. At 4 months of age, the infant developed severe respiratory failure, requiring cardiopulmonary resuscitation, and then underwent internal stenting of the left main bronchus with a 3.0-mm diameter tracheal tube. The infant was referred to our hospital for further respiratory management at 5 months of age. Chest computed tomography (CT) revealed compression of the left main bronchus by the left pulmonary artery (LPA), and rigid bronchoscopy showed complete obstruction of the left main bronchus with granulation tissue (Fig. 1a). Urgent surgical intervention was performed through a median sternotomy. The left lung crossed over into the right chest. After opening the pericardium in the midline, the main pulmonary artery and single LPA were observed just under the pericardium. Arteriopexy of the LPA (LPA-pexy) was performed with two horizontal mattress sutures of pledgeted 4-0 polypropylene placed into the anterior LPA wall. Because intraoperative flexible bronchoscopy still showed collapse of the left main bronchus, three interrupted sutures of 4-0 polypropylene were placed on the left pericardial edge adjacent to the LPA and fixed to the left hemisternum. Intraoperative bronchoscopy after the LPA-pexy with pericardial suspension confirmed adequate opening of the collapsed bronchial lumen (Fig. 1b). The postoperative course was uneventful without internal stenting of the left main bronchus. The infant was transferred back to his referring hospital on postoperative day 17 and experienced no postoperative complications during the subsequent 6 months of clinical follow-up.

Case 2

A male infant was born with a birth weight of 2515 g at 39 weeks of gestation. He was presented with stridor since birth and had repeated episodes of respiratory distress that required frequent hospitalization. Chest CT revealed right UPA and congenital tracheal stenosis (Fig. 2a). At 18 months of age, the infant developed acute respiratory failure after an upper respiratory infection and required tracheal intubation and mechanical ventilation. The infant was referred to our hospital for surgical treatment of congenital tracheal stenosis. Rigid bronchoscopy showed long-segment tracheal stenosis with complete cartilaginous rings, extending proximally from the first tracheal ring and distally 25 mm above the level of the carina. Emergency slide tracheoplasty was performed with cardiopulmonary bypass support. To prevent postoperative tracheomalacia due to direct aortic compression, the trachea was translocated anterior to the aorta, and tracheopexy was performed with pericardial suspension. Postoperative chest CT revealed good patency of the reconstructed trachea anterior to the aorta and improvement of the hyperinflation of the left lung (Fig. 2b). Although the infant underwent tracheostomy because of subglottic stenosis postoperatively, he was successfully weaned from the mechanical ventilator. The infant was transferred back to his referring hospital at 22 months of age and had an uneventful postoperative course during the 6-month follow-up.

Figure 2



Chest computed tomographic findings of case 2. (a) Preoperative image showing the stenotic trachea posterior to the aorta and hyperinflated left lung extending across the midline. (b) Postoperative image showing reconstructed trachea anterior to the aorta and reduced hyperinflation of the left lung. The dashed line indicates the direction of pericardial suspension. Ao, aorta; Tr, trachea.

Comment

Aortopexy/arteriopexy or tracheopexy/bronchopexy is considered as one of the options for surgical treatment when tracheobronchomalacia is caused by extrinsic compression in infancy. Pulling up the overlying vessel or the airway itself anteriorly to the sternum can reduce the tracheobronchial collapse and improve respiratory function. In patients with right UPA, displacement and rotation of the heart to the hemithorax have the potential to cause aortic compression of the trachea, and our previous study showed that aortopexy or tracheopexy were also useful to prevent postoperative tracheomalacia [3]. However, the anterior border of the left lung often crosses the midline because of hyperinflation, which interrupts the route to the sternum for aortopexy or tracheopexy in patients with right UPA. In addition, the hyperinflated lung may exacerbate the displacement and rotation of the heart to the hemithorax by compressing the heart, resulting in a negative impact on cardiac

function. Pericardial suspension restrains the hyperinflated lung from entering into the anterior mediastinum and creates space for aortopexy or tracheopexy [4]. It may mitigate cardiac compression of the hyperinflated lung. Recently, Jennings *et al.* [5] reported that additional traction sutures placed in the adjacent pericardium have occasionally been required for effective surgical treatment of severe tracheobronchomalacia. In case 1 described here, intraoperative bronchoscopy showed that the tracheobronchial collapse was improved more by LPA-pexy with pericardial suspension than by LPA-pexy only. The outcome in case 2 indicates that pericardial suspension concomitant with aortopexy would be more effective to prevent postoperative tracheobronchomalacia after slide tracheoplasty in patients with UPA. Although internal [6] and external stenting [7] are other therapeutic options for severe tracheobronchomalacia, we do not consider these airway stents as a first choice of treatment for a malacic lesion caused by extrinsic vascular compression. Chin *et al.* [8] reported that airway stenting is unsuitable for treating extrinsic vascular compression because of a high rate of erosion, hemorrhage, formation of obstructive granulation tissue, and airway perforation. In addition, external stenting using a foreign material may cause infection because tracheoplasty is not a completely aseptic operation [9]. A major concern as regards pericardial suspension in patients with UPA may be a reduction in ventilation volume in the single lung with compensatory hyperinflation. However, both patients described in this report had an uneventful postoperative course, with improvement in respiratory function. Another concern is that excess pericardial suspension may exacerbate cardiac function by suspension of the heart. Monitoring of arterial blood and central venous pressure is also important to decide the optimal pericardial suspension.

In summary, pericardial suspension is supportive and effective in surgical airway management of tracheobronchomalacia in patients with UPA, because this technique creates space for aortopexy or tracheopexy, mitigates cardiac compression of the hyperinflated lung, and improves tracheobronchial patency.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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