Surgical repair of pulmonary artery sling without concomitant correction of associated tracheal abnormalities has yielded poor results in the past. We combined vascular sling repair done during cardiopulmonary bypass through a median sternotomy incision with tracheopexy for severe associated tracheomalacia in 2 infants.

A

lthough vascular repair alone of pulmonary artery sling has successfully relieved respiratory symptoms [1], many patients continue to have obstructive airway symptoms that prolong postoperative recovery and increase operative mortality [2, 3]. Tracheal anomalies occur in about 50% of these cases and include localized tracheomalacia and fixed stenosis from complete tracheal rings [4]. Relief of tracheal obstruction may be difficult when segments of fixed stenosis are present, but may be more easily accomplished in patients with localized tracheomalacia by using techniques to put external traction on the involved segment as have been described for tracheomalacia with tracheoesophageal fistula [5]. We describe 2 infants with vascular sling and tracheomalacia in whom cardiopulmonary bypass was used for vascular repair and then support of the abnormal segment of trachea was achieved by modified aortopexy and tracheopexy.

Patient 1

A 3.36-kg girl with Down’s syndrome became cyanotic and dyspneic two hours after birth and required endotracheal intubation. She then improved and was extubated but underwent cardiac catheterization because of a systolic murmur. She was found to have a pulmonary artery sling with a ventricular and atrial septal defect with a pulmonary to systemic blood flow ratio of 2.3:1. She improved on medical therapy and was discharged home. After 1 week, she again had episodes of cyanosis and was readmitted. A chest roentgenogram during a cyanotic spell showed hyperinflation of the left lung.

At 7 weeks of age, operation was performed after bronchoscopy showed marked slit-like narrowing of the distal trachea, carina, and left mainstem bronchus. Using a median sternotomy incision and cardiopulmonary bypass, the ductus arteriosus was divided and a small ventricular and atrial septal defect were both closed by direct suture. The left pulmonary artery was detached and dissected from behind the trachea and distally toward the hilum to ensure a nonobstructive course after anastomosis. Anastomosis with the main pulmonary artery was performed using continuous 6-0 PDS suture posteriorly and interrupted 7-0 Prolene (Ethicon, Houston, TX) anteriorly. After discontinuing cardiopulmonary bypass, the distal trachea and carina were exposed and clearly lacked adequate cartilaginous support. The aorta was directly anterior to the involved airway, the abnormal distal trachea and left mainstem bronchus were first sutured to the adventitia of the posterior aortic wall. Before chest closure, sutures placed in the adventitia of the aorta anteriorly and then to the endothoracic fascia lateral to the sternum were tied, thus placing moderate anterior traction on the abnormal airway. Bronchoscopy immediately after chest closure showed open airways with only minor distortion of the lumen of the left mainstem bronchus.

The patient did well postoperatively, was extubated on the first postoperative day, and had no respiratory difficulties. She was discharged on the 11th postoperative day. Recatheterization at 1.5 years showed an open left pulmonary artery anastomosis with mild stenosis (6 mm Hg gradient). She continues to do well and has no respiratory symptoms.

Patient 2

This patient was admitted at 3 weeks of age with dyspnea and wheezing, treated for pneumonia, and discharged after 6 days. At 6 weeks of age, he had severe respiratory distress with wheezing and fever and was readmitted after resuscitation from a respiratory arrest. Treatment was begun for presumed bronchiolitis, but he deteriorated and required endotracheal intubation. Cardiac catheterization done because of suspected heart disease with pulmonary edema showed pulmonary artery sling and a possible small atrial septal defect. On bronchoscopy there was a narrow distal trachea from external compression and a markedly narrowed left mainstem bronchus. After resolution of his respiratory infection, he was able to be...
Fig 1-Left anterior oblique view of the establishment of external support of airway involved with tracheomalacia in patient 2.

Weaned from the ventilator, improved, and was discharged from the hospital.

At age 12 weeks he had recurrent pulmonary congestion and cough and then became cyanotic and apneic, and again required resuscitation and endotracheal intubation. At age 13 weeks, he underwent an operation using a median sternotomy and cardiopulmonary bypass. The ductus was divided and no atrial septal defect was found. The mobilization and anastomosis of the left pulmonary artery were done as described for patient 1. As the involved airway did not lie directly behind the aorta but more to the right, we placed four sutures of 5-0 Prolene in the anterior tracheal wall, carina, and proximal left mainstem bronchus, brought these directly through the intercostal spaces lateral to the sternum, and tied them with moderate tension just after sternal closure (Fig 1). Bronchoscopy after chest closure showed an open airway with only mild narrowing of the left mainstem bronchus.

Because of marked fluid retention associated with preoperative paralysis, the patient first underwent diuresis and then was extubated on the fifth postoperative day. He hereafter had normal respiratory function and was discharged on the 13th postoperative day. Cardiac catheterization performed at age 1.5 years showed a normal-sized Patent leh pulmonary artery anastomosis. He has not had significant respiratory symptoms since his operation.

Comment

Though pulmonary artery sling can be asymptomatic or have early symptoms that later resolve, patients with recurrent respiratory compromise such as ours will have a high early mortality without surgical intervention [6]. The dismal results of surgical repair of vascular sling in the past have been due to the poor patency rate of the vascular anastomosis and the persistence of respiratory compromise due to associated tracheal abnormalities. Although the left thoracotomy approach has been preferred by some surgeons [1, 3], a median sternotomy with cardiopulmonary bypass provides better exposure and, we believe, the best conditions to assure a technically perfect vascular anastomosis. Bronchoscopy before repair is essential to determine whether there is fixed stenosis or substantial localized tracheomalacia. Single-stage repair of pulmonary artery sling with tracheal resection for fixed stenosis has been successfully performed and should improve results for this combination of lesions [7]. Localized tracheomalacia can also severely compromise postoperative respiratory function and has been corrected by external support using aortopexy or tracheopexy in 2 cases reported by Campbell and colleagues, 1 done late after repair of pulmonary artery sling and 1 done concomitantly with vascular repair [8].

Our experience supports the use of single-stage repair for pulmonary artery sling using cardiopulmonary bypass for vascular repair and adding external airway support by tracheopexy for major segmental tracheomalacia. Although we performed bronchoscopy before and immediately after chest closure to confirm proper suture placement and effective airway support, with the current availability of a very small fiberoptic endoscope, it would be better to perform the tracheal suspension while visualizing the effect endoscopically. Although we describe our successful experience in only 2 patients using this approach, considering the severity of their preoperative symptoms and bronchoscopic findings, it seems justified to conclude that the tracheal procedure contributed substantially to their freedom from early postoperative respiratory difficulties.

References