Surgical Treatment of Tracheomalacia by Bronchoscopic Monitored Aortopexy in Infants and Children

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Background. Aortopexy has become an established surgical procedure for the treatment of severe tracheomalacia (TM) in infancy. However, postoperative outcome may be improved by intraoperative bronchoscopic control of the aortopexy.

Methods. Between 1992 and 2000, 16 infants and children (2 female, 14 male) with TM were treated by pexis of the aorta via a right (15 patients) or left (1 patient) anterior thoracotomy. Patients age ranged from 4 to 122 months (mean, 26 mon). Three infants had previous surgery for esophagus atresia and tracheoesophageal fistula. Another four patients were operated for gastroesophageal reflux. In all cases, the aortopexy was monitored intraoperatively by bronchoscopy. Respiratory function was verified for each patient by comparing pre- and postoperative tidal expiratory flow values (TEF 25% in ml/sec).

Results. Mean follow-up was 36 months (range, 2 to 60 mo). There was no intraoperative or postoperative mortality. 13 patients showed permanent relief of symptoms. Postoperative median TEF 25% increased significantly compared with preoperative values (81 ml/sec vs. 56 ml/sec; p = 0.016). In one patient repeat aortopexy was necessary.

Conclusions. Aortopexy through a right anterior thoracotomy is an efficient and feasible method in the surgical treatment of TM in infancy and, therefore, can improve postoperative respiratory function. Intraoperative bronchoscopy is advantageous.

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Tracheobronchomalacia (TBM) is a rare congenital or acquired disease characterized by abnormal softness or absence of the bronchotracheal cartilaginous bridges resulting in a reduced tracheobronchial lumen. Additional flaccidity may worsen the process which can involve the entire trachea or only segments of the tracheobronchial tree. In cases of primary (congenital) bronchomalacia (BM) oesophageal atresia and tracheoesophageal fistula are usually associated [1, 2]. Secondary (acquired) BM is usually caused by continuous compression due to vascular malformations, tumors, long term intubation or tracheostomy [3].

The spectrum of clinical symptoms ranges from mild and recurrent respiratory infections to severe and acute airway obstruction with apneic and cyanotic episodes or so-called dying spells. Typically, symptoms develop in early infancy but may first present even later on in childhood depending upon their severity.

Mild cases usually resolve spontaneously or under conservative treatment whereas severe forms with progressive symptoms require surgical intervention early in infancy. The technique and timing of operation remain controversial. Aortopexy, as first described by Gross and Newhauser, is accepted as a surgical procedure for the treatment of severe TM, avoiding complex tracheobronchial resections [4]. In the present study we retrospectively demonstrate our experience with bronchoscopic monitored aortopexy in 16 infants and children.

Patients and Methods

Demographic Data and Diagnoses

Sixteen infants (2 female, 14 male) with severe TM were treated by aortopexy (Table 1). Patients age ranged from 4 to 122 months (mean, 26 mon). Mean body weight was 12.9 kg (range, 3.5 to 12.0 kg). Five infants had previous surgery for esophageal atresia. In two of these 5 cases a recurrent tracheoesophageal fistula was observed. Another four patients were operatively treated for gastroesophageal reflux. One patient was a premature newborn (31 weeks of gestation) and another infant had a 21p+ chromosomal defect with choanal stenosis. In one patient a left sided thoracotomy was performed for simultaneous resection of a persistent ductus arteriosus that was responsible for stenosis of the left main bronchus.

The most frequent symptom was recurrent respiratory distress and pneumonia, with more than 3 episodes in 12 patients (75%). Nine patients (56%) had recurrent apnea and cyanotic episodes. Inspiratory stridor was observed
Table 1. Baseline Characteristics of 16 Patients With Tracheomalacia

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Sex (m/f)</th>
<th>Age (mo)</th>
<th>Body Weight (kg)</th>
<th>Symptoms</th>
<th>Comorbidity</th>
<th>Previous Operations</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>m</td>
<td>4</td>
<td>8.4</td>
<td>recurrent apnea</td>
<td>GER</td>
<td>fundoplication</td>
</tr>
<tr>
<td>2</td>
<td>m</td>
<td>6</td>
<td>7.3</td>
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<td>PDA</td>
<td>simultaneous PDA closure through left-sided thoracotomy</td>
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<tr>
<td>3</td>
<td>m</td>
<td>4</td>
<td>8.0</td>
<td>pneumonia</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>m</td>
<td>8</td>
<td>8.5</td>
<td>recurrent apnea, pneumonia</td>
<td>GER</td>
<td>fundoplication</td>
</tr>
<tr>
<td>5</td>
<td>m</td>
<td>11</td>
<td>9.0</td>
<td>recurrent apnea, pneumonia</td>
<td>GER</td>
<td>fundoplication</td>
</tr>
<tr>
<td>6</td>
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<td>8.1</td>
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<td>GER</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>m</td>
<td>6</td>
<td>7.5</td>
<td>recurrent apnea, pneumonia</td>
<td>re-tracheoesophageal fistula</td>
<td>correction of EA</td>
</tr>
<tr>
<td>8</td>
<td>m</td>
<td>5</td>
<td>5.1</td>
<td>inspiratory stridor, pneumonia, feeding problems</td>
<td>re-tracheoesophageal fistula</td>
<td>correction of EA</td>
</tr>
<tr>
<td>9</td>
<td>f</td>
<td>21</td>
<td>11.4</td>
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<td>re-tracheoesophageal fistula</td>
<td>correction of EA</td>
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<td>25</td>
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<td>correction of EA</td>
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<td>11</td>
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<td>21p+ chromosomal defect with choanal stenosis</td>
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</tr>
<tr>
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<td></td>
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<tr>
<td>13</td>
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<td>18</td>
<td>inspiratory stridor, pneumonia</td>
<td>DAA</td>
<td></td>
</tr>
</tbody>
</table>

DAA = double aortic arch; EA = esophageal atresia; EGP = esophagogastropexy; GER = gastroesophageal reflux; PDA = persistent ductus arteriosus.

in 7 infants, whereas only two of them had a congenital stridor. Life-threatening dying spells after a prolonged apneic or cyanotic period were seen in 6 patients (37%) as the most serious problem. All these infants required cardiopulmonary resuscitation. One of these patients was intubated before operation. Another infant had feeding and thriving problems.

Preoperative Bronchoscopy and Lung Function Test

TM was diagnosed preoperatively by flexible bronchoscopy (Olympics BF Type N20) in all patients breathing spontaneously. In all cases a short segment obstruction (1-3 cm) of the distal trachea was found. During expiration tracheal collapse, softness of cartilaginous bridges or an abnormally wide pars membranacea bulging into the tracheal lumen was observed most frequently. To exclude tracheoesophageal compression by aortic arch anomalies, high resolution computed tomography and/or echocardiography was done. A baby lung function test (SensorMedics 2600, Anaheim, CA, USA) was performed pre- and postoperatively [5]. In order to assess postoperative outcome, median tidal expiratory flow values (TEF 25%) were estimated and compared with corresponding values [6].

Surgical Technique and Intraoperative Bronchoscopy

Patients were positioned supine on the table with the right chest slightly elevated. In all patients access to the aorta was obtained by a right anterior thoracotomy through the 4th intercostal space (incision length, 4-7 cm). If necessary the right lobe of the thymus was resected in order to expose the ascending aorta. Avoiding injury of the right phrenic nerve, the pericardium was opened longitudinally to the aortic duplication fold. One or two 2-0 pledged nonabsorbable ethibond sutures were placed in the pericardial duplication fold and adventitia of the ascending aorta without entering the aortic lumen (Fig 1A). After passing through the posterior periosteum of the sternum the sutures were suspended forward to bring the aortic root to the sternum. Confirming the most effective direction by intraoperative bronchoscopy, the sutures were then tied (Fig 1B). In Figures 2A and B the corresponding bronchoscopic views of tracheal stenosis before and its decompression immediately after aortopexy are presented. Postoperatively, all patients were treated with inhalative steroids to reduce tracheobronchial hyperreactivity.

Statistics

Statistical analysis of the data was performed using the "StatView" software package (Abacus Concepts, Berkeley, CA, USA). A t-test was applied to compare intradividual values. The significance level was established at $a = 0.05$.

Results

All 16 infants underwent aortopexy without intra- or postoperative mortality. In one case the sutures tore out so that repeat aortopexy three months after the initial
procedure was necessary. In all cases extubation could be performed immediately after operation in the operation theatre. Permanent relief of symptoms (free of stridor, no further apnic episodes) was clinically observed in 13 patients. Mean follow-up was 36 months, ranging from 2 to 60 months. Another patient presented a relapse of tracheoesophageal fistula three years after aortopexy which was successfully repaired by bronchoscopic closure. In patients who had a preoperative lung function test, the median TEF 25% was significantly increased after aortopexy (81 ml/sec vs. 56 ml/sec; $p = 0.016$) (Fig 3).

**Comment**

Tracheomalacia that occurs as a primary condition in the absence of other lesions may be due to a developmental defect of cartilaginous rings [2]. It is well known that tracheomalacia and bronchomalacia can also be acquired. This occurs mainly in the population of patients with chronic obstructive lung disease. Persistent high airway pressures may be responsible for ongoing destruction of tracheal and bronchial cartilage as well as enlarged major airways with tendency to collapse. When it is found in association with tracheal collapse, it may be difficult to determine which mechanism is responsible for the clinical symptoms. In fact, gastroesophageal reflux may cause tracheal collapse if the upper oesophagus enlarges during regurgitation and impinges on a soft trachea [7].

Aortopexy has been proven to be a safe and simple method for the management of severe TM in infancy [8, 9, 10]. It does not alter the tracheal structure and works by pulling the trachea forward, thus widening tracheal diameter. However, the operative technique is still controversial. Many authors preferred a left anterior thoracotomy in the 3rd intercostal space and reported good
exposure of the ascending aorta and the branches of the aortic arch [7, 11, 12]. Brawn et al. described successful tracheoaortopexy via a midline sternotomy in two infants with proximal tracheomalacia [13]. An anterior cervical approach for tracheopexy was presented by Vaishnav et al. as alternative [14]. We prefer a right anterior thoracotomy which allows adequate exposure of the aorta and access to the innominate artery. Bullard and co-workers described a mediastinal window approach by using a small transverse incision over the 3rd intercostal space for aortopexy avoiding a standard thoracotomy [15]. A dacron patch aortopexy is favoured by Spitz to minimize the aortic trauma [16]. Applebaum et al. modify the standard suture aortopexy by using a pericardial flap as a tough structure for suspending the aorta without sutures placed in the aortic wall [17].

However, there are still some open questions regarding optimal treatment of TM in infancy. Based on our experience, 70% of the cases with TM did not require aortopexy because symptoms resolved spontaneously. In 30% of the patients presenting severe symptoms with life-threatening episodes aortopexy was indicated. All these patients had a TM based on a short obstruction of the distal trachea. Aortopexy might not be sufficient in the treatment of airway collapse when the entire length of intrathoracic trachea was involved. Endotracheal or external stenting techniques were reported to be feasible for reinforcing the tracheal wall, thus preventing airway collapse [18,19]. Airway splinting in management of segmental TM did not seem to affect tracheal growing either experimentally or in humans [20, 21]. However, tracheostomy should be avoided as a primary approach since secondary TM and tracheal fibrosis may occur [3].

According to our experience, early aortopexy can be recommended in infants with severe symptoms and short segment obstruction in the distal trachea with severe symptoms. Intraoperative bronchoscopic monitoring should be performed to control the amount and direction of aortopexy assuring the most effective tracheal decompression [22].

References
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Southern Thoracic Surgical Association: Forty-ninth Annual Meeting

The Forty-ninth Annual Meeting of the Southern Thoracic Surgical Association will be held November 7-9, 2002, Miami Beach, Florida.

The Postgraduate Course will be held the morning of Thursday, November 7, 2002, and will provide in-depth coverage of thoracic surgical topics selected primarily as a means to enhance and broaden the knowledge of practicing thoracic and cardiac surgeons.

Manuscripts accepted for the Resident Competition need to be submitted to the STSA headquarters office no later than September 13, 2002. The Resident Award will be based on abstract, presentation, and manuscript.

Applications for membership should be completed by September 15, 2002, and forwarded to Geoffrey M. Graeber, MD, Membership Committee Chairman, Southern Thoracic Surgical Association, 401 N Michigan Ave, Chicago, IL 60611-4267.

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