Resection of the Right Middle Lobe and Lingula in Children for Middle Lobe/Lingula Syndrome*

Adel K. Ayed, MD, FCCP

**Study objectives:** To review our experience with specific characteristics, indications, and results of pulmonary resection in children with middle lobe/lingula syndrome.

**Design:** Retrospective cohort study.

**Setting:** Thoracic Surgery Department, Chest Diseases Hospital, Kuwait.

**Patients and intervention:** Thirteen children with middle lobe, lingula, or both syndromes were treated with pulmonary resection from January 1995 to December 1999.

**Results:** The mean age was 7.5 years (range, 5 to 10 years). Eight patients were girls, and five were boys. All patients underwent high-resolution CT and bronchoscopy. Bronchiectasis and atelectasis of right middle lobe, lingula, or both was noted in nine patients. Bronchial stenosis and inflammation of the bronchus was found endoscopically in four patients. The indications for surgery were recurrent respiratory tract infection with persistent atelectasis and bronchiectasis in nine patients, and recurrent respiratory tract infection with bronchiectasis in four patients. A right middle lobectomy was done on seven patients and a lingulectomy on four patients. Two patients underwent staged thoracotomies (right middle lobectomy and lingulectomy). There were no operative deaths. Only two patients had postoperative complications: atelectasis (n = 1), and pneumothorax (n = 1). Mean follow-up was 3.5 years (range, 3 to 5 years) for all patients. Nine patients were asymptomatic, and four patients had improved.

**Conclusion:** Right middle lobe or lingula syndrome with the presence of bronchiectasis, bronchial stenosis, or failure of lung to re-expand are indications for early pulmonary resection.

*(CHEST 2004; 125:38–42)*

**Key words:** atelectasis; bronchiectasis; pulmonary resection; right middle lobe

**Abbreviations:** HRCT = high-resolution CT; RMLS = right middle lobe syndrome

---

From a large group of patients with bronchiectasis, a subgroup has been noted who have involvement of the right middle lobe, lingula, or both. Right middle lobe and lingular bronchiectasis has long been recognized as a unique clinical syndrome.1,2 Right middle lobe syndrome (RMLS) is characterized by a spectrum of diseases from recurrent atelectasis and pneumonitis to bronchiectasis of the middle lobe.2 It has been described among all age groups.2 The mechanisms of RMLS in children were thought to be a nonobstructive atelectasis due to poor collateral ventilation.1,3,4 A specific group of these patients were examined. The purpose of this study was to analyze the specific characteristics of patients with bronchiectasis of the middle lobe, lingula, or both, and to document the indications and results of surgical resection.

**Materials and Methods**

A retrospective review of 13 children < 15 years old with atelectasis and bronchiectasis of the right middle lobe, lingula, or both was undertaken at the Chest Diseases Hospital between January 1995 and December 1999. The data obtained included age, sex, presentation, location of the lesion, method of workup, and the operative treatment. Follow-up was regular in the outpatient department every month for 3 months, and every 3 months thereafter for 3 to 5 years. Follow-up included clinical history, physical examination, chest radiography.

**Results**

Seven children were operated on for right middle lobe disease (Figs 1, 2), four children were operated...
on for lingular disease (Fig 3), and the remaining two children had both the middle lobe and lingula resected for bilateral disease (Table 1). Age at the time of operation ranged from 5 to 10 years. Eight patients were girls, and five were boys. The presenting symptoms were recurrent respiratory tract infections in all patients. The periods of symptoms are reported in Table 1.

All patients underwent preoperative high-resolution CT (HRCT) to evaluate the severity and distribution of bronchiectasis. Sputum was cultured for bacterial examination before operation. All patients were screened by bronchoscopy to exclude causes such as a central obstruction or stenosis of bronchus caused by a foreign body, tumor, or other cause. As a result, the bronchial stenosis was discovered in four cases. One patient had a granulation tissue partially
<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Year of Operation</th>
<th>Age, yr</th>
<th>Sex</th>
<th>Period Between Onset of Symptoms and Surgery</th>
<th>Area Resected</th>
<th>Lung Function, % Predicted</th>
<th>Bronchoscopy</th>
<th>CT Scan</th>
<th>Postoperative Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1995</td>
<td>8</td>
<td>Female</td>
<td>6 mo</td>
<td>RML</td>
<td>Preoperative 71, Postoperative 83</td>
<td>Stenosis of RML bronchus</td>
<td>Atelectasis and bronchiectasis</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>1995</td>
<td>5</td>
<td>Female</td>
<td>1 yr</td>
<td>RML</td>
<td>Preoperative 90, Postoperative 88</td>
<td>Secretions</td>
<td>Atelectasis and bronchiectasis</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>1995</td>
<td>10</td>
<td>Female</td>
<td>1 yr</td>
<td>Lingula</td>
<td>Preoperative 100, Postoperative 96</td>
<td>Bronchiectasis</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>1996</td>
<td>8</td>
<td>Male</td>
<td>8 mo</td>
<td>RML</td>
<td>Preoperative 103, Postoperative 100</td>
<td>Secretions</td>
<td>Bronchiectasis</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>1996</td>
<td>8</td>
<td>Female</td>
<td>6 mo</td>
<td>Lingula</td>
<td>Preoperative 85, Postoperative 76</td>
<td>Secretions</td>
<td>Bronchiectasis</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>1996</td>
<td>10</td>
<td>Male</td>
<td>5 mo</td>
<td>RML</td>
<td>Preoperative 82, Postoperative 94</td>
<td>Granulation tissue RML</td>
<td></td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>1997</td>
<td>7</td>
<td>Female</td>
<td>7 mo</td>
<td>RML</td>
<td>Preoperative 77, Postoperative 72</td>
<td>Secretions</td>
<td>Atelectasis and bronchiectasis</td>
<td></td>
</tr>
<tr>
<td>8</td>
<td>1998</td>
<td>5</td>
<td>Female</td>
<td>8 mo</td>
<td>Lingula</td>
<td>Preoperative 96, Postoperative 94</td>
<td>Secretions</td>
<td>Atelectasis and bronchiectasis</td>
<td></td>
</tr>
<tr>
<td>9</td>
<td>1998</td>
<td>10</td>
<td>Female</td>
<td>1 yr, 3 mo</td>
<td>RML</td>
<td>Preoperative 96, Postoperative 94</td>
<td>Stenosis of RML bronchus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>1999</td>
<td>6</td>
<td>Female</td>
<td>10 mo</td>
<td>Lingula</td>
<td>Preoperative 77, Postoperative 72</td>
<td>Secretions</td>
<td>Atelectasis and bronchiectasis</td>
<td></td>
</tr>
<tr>
<td>11</td>
<td>1999</td>
<td>7</td>
<td>Male</td>
<td>8 mo</td>
<td>Lingula</td>
<td>Preoperative 96, Postoperative 94</td>
<td>Secretions</td>
<td>Atelectasis and bronchiectasis</td>
<td></td>
</tr>
<tr>
<td>12</td>
<td>1999</td>
<td>8</td>
<td>Male</td>
<td>1 yr, 2 mo</td>
<td>RML</td>
<td>Preoperative 96, Postoperative 94</td>
<td>Stenosis of RML bronchus</td>
<td></td>
<td></td>
</tr>
<tr>
<td>13</td>
<td>1999</td>
<td>6</td>
<td>Male</td>
<td>9 mo</td>
<td>RML</td>
<td>Preoperative 77, Postoperative 72</td>
<td>Stenosis of RML bronchus</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*RML = right middle lobe.
obstructing the right middle lobe bronchus. The remaining patients had secretions in the bronchus. No patients had a foreign body. Bacteria was preoperatively proven in 11 patients: Haemophilus influenzae in 5 patients, Pseudomonas aeruginosa in 3 patients, Staphylococcus aureus in 2 patients, and Streptococcus pneumoniae in 1 patient. Sputum was negative for acid-fast bacilli in all patients. All patients had negative screening test results for cystic fibrosis and immune deficiency disease. Pulmonary function test results in some patients are reported in Table 1.

All 13 patients had been receiving regular treatment with bronchodilators, mucolytic agents, antibiotics, chest physiotherapy, and postural drainage for long periods, prior to surgical resection. The decision to operate was based on the chronicity of the disease, the presence of severe bronchiectasis, or an entirely destroyed lobe or segment. Right middle lobectomy was performed on seven patients and lingullectomy on four patients. Two patients underwent staged thoracotomies (right middle lobe and lingullectomy). There was no operative mortality within 30 days. Complications occurred in two patients (15%), including atelectasis (requiring bronchoscopy in one patient), and pneumothorax (requiring reinsertion of a chest tube in one patient). Ten patients were discharged 6 days after surgery; the remaining 3 patients were discharged at 8 days, 9 days, and 12 days, respectively. Postoperative care was standard as in most pulmonary resections, and included early mobilization and aggressive chest physiotherapy.

All patients were followed up regularly in the outpatient department for a mean of 3.5 years (range, 3 to 5 years). All patients were either completely well or had fewer pulmonary symptoms postoperatively than preoperatively. Of the 11 patients who received a one-side operation, 8 patients have been asymptomatic since surgery and have no evidence of disease in the opposite side. The remaining three patients have had recurrence of their symptoms 6 months postoperatively and evidence of mild bronchiectasis in the opposite side. Their symptoms included mild cough and sputum production and required mucolytic agents and occasional antibiotics. One patient who received bilateral surgery had hyperactive airway disease and required bronchodilators and occasional steroids.

DISCUSSION

RMLS is characterized by a spectrum of diseases from recurrent atelectasis and pneumonitis to bronchiectasis of the middle lobe. It has been described among all age groups, although the diagnosis in pediatrics may be delayed or missed because of nonspecific symptoms or findings.\(^1\)\(^2\)

The causes of middle lobe syndrome may be obstructive, extraluminal or intraluminal, or nonobstructive. Extraluminal obstruction can be due to bronchial compression by tumor or lymph nodes, abnormal branching, or abnormal bronchial diameter, length, or structure.\(^3\) Aspiration of foreign material, edema or scarring of the mucosa, and granulation tissue or tumors cause intraluminal obstruction.\(^3\) Primary inflammatory processes and disorders of collateral ventilation are designated as nonobstructive sources of atelectasis syndrome.\(^2\)\(^–\)\(^4\) The middle lobe and lingula contain only scanty parenchymal bridges due to the deep fissures that isolate these lobes and provide effective barriers to collateral ventilation, thus predisposing to chronic inflammation and persistent atelectasis.\(^3\)\(^4\)

Our experience indicates that the middle lobe is prone to develop persistent total or partial atelectasis with infection. Nine patients in this series had atelectasis and bronchiectasis, and infection was a manifestation of the syndrome in all patients. However, with repeated episodes of infection, through the resulting vicious cycle of recurring bouts of inflammation, the right middle lobe may eventually be destroyed completely.\(^1\) The etiology of infection is commonly bacterial and occasionally viral, and in some cases it is caused by tuberculosis.\(^5\)\(^6\) Eleven patients in this series had bacterial infections, and none had tuberculosis. The diagnosis was made by culture of the bronchial aspirates. Allergy has long been known to cause middle lobe syndrome.\(^2\) Edema of the bronchial wall, bronchospasm, and increased production of mucus have been considered as precipitating factors. In this series, none of the patients had allergy or asthma. One patient had hyperactive airway disease on follow-up and required bronchodilators with occasional steroids.

The diagnosis of middle lobe/lingula syndrome is made by clinical and radiographic examination of the chest.\(^7\)\(^8\) A history of recurrent pneumonia, fever, productive cough, and wheezing is obtained in the majority of patients. Chest radiographic findings are suggestive of diagnosis when both posteroanterior and lateral views are obtained. The classic finding is on the lateral view where atelectatic middle lobe is seen as wedge-shaped density extending from hilum anteriorly and inferiorly (Fig 1). In this series, such a finding was noted in only nine patients. Four patients had bronchiectasis of the middle lobe or lingula or both. HRCT is currently a modality of choice in the diagnosis of bronchiectasis, with only a 2% false-negative rate and 1% false-positive rate.\(^9\) HRCT was done in all patients in this series to document the presence of bronchiectasis. Bronchosc-
copy is important for diagnosis and treatment. The status of the orifice and proximal portion of the middle lobe/lingula bronchus can be visualized using bronchoscopy. Bronchial obstruction by granulation tissue, tumor, or by a foreign body can be ruled out with a great certainty. In our series, we found four patients who had stenosis and inflammation of the bronchus. Bronchoscopy is also helpful in the removal of retained secretions and allows to take bronchial lavage for microbiology.

The management of middle lobe/lingula syndrome in pediatric patients has been conservative and includes the following: antibiotics, postural drainage, and bronchodilators. This intensive medical therapy is quite capable of resolving atelectasis and healing the patient. If the atelectasis has not regressed within 1 month, however, bronchoscopy and a CT scan should be carried out. If the bronchoscopy and endoscopic findings are normal and there is no evidence of bronchiectasis on CT scan, conservative treatment should be continued for a further month. If the middle lobe or lingula fails to re-expand, or recurrent atelectasis is observed or when bronchiectasis is documented, surgical excision of the right middle lobe or lingula is indicated. Operative treatment also should be offered for patients with complete bronchial obstruction. Saha et al reported 31 of 98 patients with middle lobe syndrome (32%) required surgical resection for persistent disease.

In a 10-year follow-up study of 21 children with middle lobe syndrome, Livingston et al reported a cure rate of 33% on conservative treatment, with bronchoscopy playing an important therapeutic role. In 33% of the patients, surgical resection was necessary, and the fate of the remaining third is as yet uncertain. In the study of Boeck and colleagues, the outcome of 17 children with middle lobe syndrome was evaluated. Five of the 17 study group children had ongoing respiratory problems, chest radiographic findings were abnormal in 6 patients, and pulmonary function test results were significantly lower in patients with ongoing respiratory symptoms than in the control children. In our series after pulmonary resection, patients were either cured or had a decreased number of pulmonary infections.

**Conclusion**

Pulmonary resection is indicated early in patients with isolated bronchiectasis or bronchial stenosis or recurrent atelectasis of the middle lobe, lingula, or both, before other portions of the lung become grossly diseased and are more likely to be susceptible to subsequent infection. These patients do better with surgical intervention.

**References**