The long-term pulmonary consequences of right middle lobe syndrome (RMLS) in childhood are not known. Therefore, outcome was evaluated in 17 children with RMLS diagnosed in early childhood (mean age, 3.3 years; SD, 1.1 year). Mean age at follow-up was 10.1 years (SD, 2.6 years). RMLS was defined as atelectasis of the right middle lobe (RML) of at least 1 month’s duration and visible on the lateral view of the chest radiograph as a wedge-shaped density extending from the hilum anteriorly and downward. Seventeen children without personal history of allergy or respiratory tract disease were studied as control group. Five of 17 study group children had ongoing respiratory problems: symptoms of asthma were present in 4 patients, and cylindrical bronchiectasis was present in one patient. Chest radiograph at follow-up was abnormal in six patients. Pulmonary function tests, including mean and SEM for vital capacity (VC) (82% of predicted ±7 vs 94% predicted ±3), FEV1 (77% of predicted ±12 vs 96% of predicted ±4) and their ratio (75±5 vs 85±3) were significantly lower in patients with ongoing respiratory symptoms than in the control children. The provocative dose causing a 20% decrease in FEV1 (PD20) of methacholine was significantly lower in patients with ongoing symptoms at follow-up than in control children and in patients without symptoms at follow-up (2.8 [2.2 to 3.1] vs 4.5 [2.2 to 8.8] and 9.2 [2.3 to 24] mg/mL; median and P25-75, p<0.05). Age at initial diagnosis tended to be younger in patients with ongoing symptoms at follow-up (2.3±0.7 years vs 3.8±0.4 years; p<0.08).

(CHEST 1995; 108:150-52)

PD20=provocative dose causing 20% decrease in forced expiratory volume in 1 s; RML=right middle lobe; RMLS=right middle lobe syndrome; RV=residual volume; TLC=total lung capacity; VC=vital capacity

Key words: atelectasis; childhood; pulmonary function; right middle lobe

The right middle lobe syndrome (RMLS) is characterized by recurrent or chronic atelectasis of the right middle lobe (RML). In children, the diagnosis may be delayed because of nonspecific symptoms. The etiology is not completely understood. Infection and inefficient collateral ventilation to the RML are thought to play a role. The disease may be associated with asthma and other allergic disorders.

The long-term pulmonary consequences of RMLS in childhood are not known because information on outcome is lacking. We therefore evaluated respiratory morbidity, pulmonary function tests, and chest radiographs in later childhood in a group of patients in whom RMS had been diagnosed earlier.

METHODS

The medical records of all patients with RMLS, diagnosed between 1980 and 1990 at the Department of Pediatrics, University Hospital of Leuven, were reviewed. RMLS was defined as atelectasis of the RML of at least 1 month’s duration and visible on the lateral view of the chest radiograph as a wedge-shaped density extending from the hilum anteriorly and downward. Twenty-one patients with adequately documented chronic or recurrent collapse of the RML were identified. Seventeen of these 21 patients participated in the follow-up study. There were nine boys and eight girls. Mean age at the follow-up was 10.1 years (SD 2.6 years); mean age at the time of initial diagnosis was 3.3 years (SD 1.1 year). Fourteen of the 17 patients had had two to five episodes of RML collapse. Three patients had had one prolonged episode of RML collapse, lasting 4, 6, and 9 months, respectively. In five patients, atelectasis or streaky densities in the lingula were present in addition to RML collapse. All patients had been treated conservatively. Long-term (lasting longer than 1 month) antiasthmatic therapy had been prescribed in all patients, bronchodilators in eight patients, anti-inflammatory asthma drugs in nine patients, chest physiotherapy in eight patients, and mucolytic drugs in four patients. No patient had undergone lobectomy.

The follow-up study consisted of a detailed history, physical examination, pulmonary function tests, and chest radiograph. For the purpose of the study, current respiratory symptoms were defined as episodes of wheezing, bronchitis (cough and constitutional disturbance lasting more than 5 days), or pneumonia in the 12 months before review. Atopy was defined as atopic dermatitis or serum IgE above the upper limit for age or positive skin prick test or radioallergosorbent test (RAST) to at least one inhaled allergen.

All pulmonary function tests were performed by the same physician and at least 14 days away from an episode of upper respiratory tract infection. Static lung volumes and three maximal expiratory flow volume curves were measured on a dry sealed spirometer (Morgan) attached to a helium analyzer. The curve with the largest sum of FVC plus FEV1 was examined. The results were compared with the normal values for sex and height.

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according to Zapletal.\textsuperscript{4} Methacholine provocation test was performed according to Cockcroft et al.\textsuperscript{3} Seventeen children without a personal history of respiratory tract disease or atopy served as control for the pulmonary function results.

Standard anteroposterior and lateral chest radiographs were taken and were read by a pediatric radiologist.

Statistical analysis was done using Student’s \( t \) test or Mann-Witney \( U \) test as appropriate; \( p \) values below 0.05 were considered statistically significant.

\textbf{RESULTS}

At follow-up examination, mean age was 10.1 years (SD 2.6 years). Mean interval since initial diagnosis was 6.2 years (SD 2.0 years). Mean age of children in the control group was 10.5 years (SD 2.6 years).

Five of 17 patients had had further respiratory symptoms. Four children, 6 to 10 years of age, had intermittent wheezing and shortness of breath, diagnosed as asthma, and were using antiasthma medication (salbutamol [albuterol] by metered-dose inhaler, cromolyn sodium, and theophylline). They were classified as having mild to moderate asthma. One 13-year-old girl suffered from recurrent episodes of cough with sputum production, at times associated with fever. Antibiotics, chest physiotherapy, and antiasthma drugs (budesonide [Pulmicort], salbutamol [albuterol]) were used regularly. Patients symptomatic at follow-up tended to be younger at the initial diagnosis of RMLS (2.5 ± 0.7 years) than patients asymptomatic at follow-up (3.8 ± 0.4 years; \( p<0.08 \)). Initial treatments were not significantly different between symptomatic and asymptomatic study patients. Atopic children with RMLS were not at increased risk for ongoing symptoms when compared with nonatopic children: 2 of the 5 symptomatic patients were atopic and 5 of the 12 asymptomatic patients were atopic.

Pulmonary function results in the study and control groups are reported in Table 1. Patients with ongoing symptoms of respiratory tract disease had significantly lower VC, FEV\(_1\), FEV\(_1\)/FVC ratio, and provocative dose causing 20% decrease in FEV\(_1\) (PD20) of methacholine when compared with the control group (\( p<0.05 \); unpaired Student’s \( t \) test and Mann-Whitney \( U \) test). Chest radiograph at follow-up was normal in 11 patients. Streaky densities in the RML without wedge-shaped atelectasis on lateral chest radiograph were present in five patients. Prominent bronchial markings in the RML, streaky densities, and parallel linear markings (tram track sign) suggestive of bronchiectasis in the lingula were present in one patient. Three of the five patients with ongoing respiratory symptoms had chest radiographic abnormalities.

Of the three patients with one prolonged episode of RMLS collapse, only one had ongoing symptoms of asthma and pulmonary function test result abnormalities. These three patients’ radiographs were normal at follow-up.

\textbf{DISCUSSION}

In the present study, about one third of the patients with RMLS in early childhood continue to have respiratory symptoms in later childhood, most commonly symptoms of asthma. Results of pulmonary function tests in symptomatic patients at follow-up were significantly different from those of control children and are consistent with mild obstructive airway disease and hyperreactive airway disease. Results of pulmonary function tests in patients asymptomatic at follow-up were not significantly different from those of control children. At follow-up, symptoms suggestive of chronic lower respiratory tract infection were rare and occurred in only one patient. Cylindrical bronchiectasis in the lingula was documented in this patient. Cystic fibrosis, humoral and cellular immunodeficiency, complement deficiency, and primary ciliary dyskinesia had been or were subsequently ruled out. In addition to supplicative airway disease, she has hyperreactive airway disease as measured by positive methacholine test (PD20 at 4 mg/mL) and more than 10% improvement of FEV\(_1\) after inhalation of salbutamol.

It is unclear whether pulmonary function abnormalities are a result of ongoing respiratory symptoms or vice versa or whether they both are secondary to an underlying hyperreactive airway disease. The latter, documented by the lower PD20 methacholine in symptomatic patients, seems the most likely explanation. The age of the patients at initial presentation is low (mean age, 3.3 years; SD, 1.1 year). This is in agreement with a recent study from Springer et al.\textsuperscript{2} In that study, 19 of 21 patients were younger than

\begin{table}
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\begin{tabular}{|c|c|c|c|}
\hline
& \textbf{Symptomatic Group} & \textbf{Asymptomatic Group} & \textbf{Control Group} \\
& \textbf{\( n=5 \)} & \textbf{\( n=12 \)} & \textbf{\( n=17 \)} \\
\hline
TLC, \% pred & 90 (9) & 93 (2) & 96 (3) \\
VC, \% pred & 82 (7)\textsuperscript{1} & 93 (2) & 94 (3) \\
FEV\(_1\), \% pred & 77 (12)\textsuperscript{1} & 90 (3) & 96 (4) \\
FEV\(_1\)/FVC ratio & 75 (5)\textsuperscript{1} & 80 (2) & 85 (3) \\
RV/TLC, \% pred & 118 (11)\textsuperscript{1} & 100 (10) & 98 (5) \\
PD20, mg/mL & 2.8 (2.2-3.1)\textsuperscript{1} & 9.2 (2.3-24)\textsuperscript{1} & 4.5 (2.2-8.8) \\
\hline
\end{tabular}
\caption{Pulmonary Function Test Results in Study Group Patients With and Without Ongoing Respiratory Tract Disease and in Control Children*}
\textsuperscript{*}Values are mean and SEM.
\textsuperscript{1}\( p<0.05 \) comparison between symptomatic study group and control group.
\textsuperscript{2}\( p<0.01 \) between symptomatic and asymptomatic study group; Student’s \( t \) test for all but PD20 for which Mann-Whitney \( U \) test and median and P25-P75 are used.
\end{table}
4 years of age. Probably the small diameter of the RML bronchus in young children makes the RML with its inefficient collateral ventilation very prone to total collapse, especially in a situation in which edema and bronchospasm are present. This could explain why RMLS occurs less frequently as the children grow older and the diameter of the RML bronchus enlarges.

The control group was established to compare pulmonary function test results. The lack of a proven normal chest radiograph in these patients may constitute a problem since the study patients were recruited on the basis of an abnormal chest radiograph. However, an abnormal chest radiograph is unlikely to be found in totally asymptomatic children. In addition, the unnecessary exposure of these children to radiation would pose an ethical problem.

In the present study, none of patients had been treated surgically. The rather mild abnormalities that were observed after RMLS during later childhood are indeed further symptoms of conservative management in most patients with RMLS. Surgical excision of the RML is suggested when bronchiectasis is documented, when atelectasis is associated with systemic symptoms, or when chronic infection poses a threat to the remainder of the lung. With the advent of newer diagnostic techniques such as fiberoptic bronchoscopy and newer and more potent antibiotics, it is unlikely that these older recommendations are still applicable nowadays.

In summary, after RMLS in early childhood, ongoing symptoms of respiratory tract disease occur in about one-third of the patients. Chronic suppurative airway disease after RMLS is rare. Vital capacity, FEV₁, and FEV₁/FVC ratio in children symptomatic at follow-up are significantly worse than in control children. Results of pulmonary function tests in children asymptomatic at follow-up are comparable to results of pulmonary function tests in control children.

REFERENCES