After surgery the patient continued to have mild respiratory distress and right pulmonary hyperinflation. Therefore, his aortic suspension was revised through a median sternotomy one month later. Capillary blood gas levels at the time of discharge showed a pH of 7.41, a carbon dioxide tension (Pco₂) of 47 mm Hg, and an oxygen pressure (Po₂) of 41 mm Hg. A chest x-ray film at the time of discharge showed return of the mediastinum to the normal position and absence of hyperinflation (Fig 3). The patient has done well since his reoperation. Respiratory distress completely resolved, and weight improved. One year following the patient's aortectomy, a left modified Blalock-Taussig shunt was performed uneventfully through a left thoracotomy.

**DISCUSSION**

Right aortic arch is widely recognized as a cause of obstruction of the airway when it forms part of a vascular ring around the trachea. Several isolated aortic arch anomalies in the absence of a vascular ring may also produce obstruction of the airway, including an anomalous origin of the left innominate artery and a normal left aortic arch displaced rightward secondary to a hypoplastic right lung. Edwards has reported postmortem findings in a patient with tetralogy of Fallot, right aortic arch, and aberrant left subclavian artery whose right main-stem bronchus was compressed by a large right aortic arch.

In our patient, progressive bronchomalacia and tracheomalacia was caused by a large right aortic arch compressing the right main-stem bronchus and trachea anteriorly. Air could enter the right lung during inspiration but could not be completely expired. This led to progressive hyperinflation of the right lung and severe respiratory decompensation with increasing cyanosis by the age of two months.

Suspension of the aorta by apposition to the underside of the sternum has been used to correct both innominate arterial and aortic compression of the trachea in infants with respiratory distress secondary to a crowded mediastinum. Because the planes of tissue surrounding the aorta, trachea, and bronchus are intact, the suspension helps to maintain patency of the structurally weakened airway. Apposition to the sternum is important for good healing. The first suspension through a right thoracotomy pulled loose. A second suspension with better exposure of the arch through a median sternotomy has remained intact and effective at 16 months of follow-up. Cardiac catheterization will be necessary to outline the anatomy of the great vessels so that future corrective surgery can be carefully planned.

In summary, this report documents for the first time severe obstruction of the airway produced by a dilated right aortic arch in association with tetralogy of Fallot and its successful surgical management. Increasing cyanosis in patients with tetralogy of Fallot may have a respiratory cause which must be differentiated from progressive hypoxemia due to decreasing pulmonary blood flow.

**REFERENCES**


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**Bronchiolitis in Rheumatoid Arthritis**

Aarne Lahdensuo, M.D., F.C.C.P.; Jorma Mattila, M.D.; and Anni Vilppula, M.D.

Bronchiolitis in association with rheumatoid arthritis has been reported, to our knowledge, in 18 patients to date. In some cases use of penicillamine has been strongly associated with the development of bronchiolitis. Most of the reported cases are described as having marked irreversible airways obstruction and hyperinflation. We describe a patient with rheumatoid arthritis whose respiratory tract symptoms began during gold therapy. Physiologic studies showed marked lung hyperinflation without pathologic findings in forced dry spirometric study. On open lung biopsy a mild degree of granulomatous bronchiolitis was found. Immunofluorescent microscopy showed IgM- and IgG-containing plasma cells in the bronchiolar walls.

Bronchiolitis obliterans has been described as a rare pleuropulmonary complication of rheumatoid arthritis. Most of the reported cases have been found to have marked irreversible airways obstruction and a fulminating course of the disease. Histology of the lung has revealed fibrous narrowing and obliteration of the bronchioles and the smallest bronchi, with infiltration of mononuclear cells. We recently encountered a patient with rheumatoid arthritis who also had granulomatous bronchiolitis in her lungs. Physiologic studies showed marked lung hyperinflation without pathologic findings in forced dry spirometric testing, and the course of the disease showed some reversible features.

**CASE REPORT**

The patient was a 44-year-old, nonsmoking woman whose grandmother had rheumatoid arthritis. She had never been exposed to any specific dusts. In 1971 she suffered from recurrent vaginal discharge, cervicitis, and hematuria due to Escherichia coli-bacterial cystitis. She had had no other previous diseases. She fell ill in 1974 with recurrence of pain in the shoulders. In 1975 she developed a seropositive rheumatoid arthritis which fulfilled the criteria for the classic class of the American Rheumatism Association. Since 1979 her arthritis has progressed continuously. During the years 1975-76, the patient was treated with penicillamine, to which her condition seemed to respond. Upon reactivation of the disease, gold therapy was started in June 1979, and she received 1,620 mg of gold altogether. In January 1980 there was a short pause in the gold therapy because of a rash. In January 1981 the patient noticed

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Chest 85/5/May 1984 705
Bronchiolitis performed.

reinstituted, with low-dose corticosteroids because of the activity of rheumatoid arthritis. In February 1982, an open lung biopsy was performed. The walls of several respiratory bronchioles were densely infiltrated with lymphocytes and histiocytes, with a small number of plasma cells and a few eosinophils (Fig 1). In the bronchiolar wall there were collections of histiocytes forming small granulomatous foci (Fig 2 and 3). The bronchiolar lumen contained mucous with polymorphonuclear leukocytes and macrophages. There was no evidence of more bronchiolar scarring, obliteration, or intraluminal polyps. In patches in the interstitium there were foci of inflammatory cells composed of lymphocytes and histiocytes. A few plasma cells were also seen. Sometimes the infiltrate was seen to extend into adjacent alveoli. Most of the alveolar spaces were, however, normal. The histiocytes formed granulomatous areas with multinucleated giant cells. Occasionally in peripheral alveolar spaces and in the adjacent interstitium there were small collections of foamy histiocytes. Foci of developing interstitial fibrosis were also seen; in these areas the alveolar lining cells were cuboidal with prominent nuclei. The pulmonary vessels were normal. For immunofluorescent microscopy, the lung biopsy was stained by the direct technique with commercially prepared fluorescein isothiocyanate-conjugated antisera to IgA, IgG, IgM, and C3. IgM- and IgG-containing plasma cells were seen in the bronchiolar walls (Fig 4). Additionally, a weak focal fluorescence of IgM was seen in some alveolar walls.

In April 1982, 30 mg/day of prednisolone was given for one month with no significant changes in respiratory symptoms or lung function results. In the middle of November 1982, the penicillamine treatment was stopped, and the patient's symptoms diminished. The lung function parameters improved in December 1982.

Discussion

Bronchiolitis in association with rheumatoid arthritis has been reported in 18 cases to date. In 1977, Geddes and co-

Figure 1. Mononuclear cell infiltration in the wall of a respiratory bronchioles. The lumen contains mucus with polymorphonuclear leukocytes and macrophages (H&E, × 225).

Figure 2. A granulomatous collection of histiocytes in the wall of a respiratory bronchioles. (H&E, × 360).

Figure 3. The interstitium contains focal infiltrates of mononuclear cells. A granulomatous focus is also seen. (H&E, × 90).

Figure 4. IgM-containing plasma cells in a bronchiolar wall. In the left upper corner a mucous plug in the bronchiolar lumen can be seen.
Workers described five patients with signs of rapidly progressive bronchiolitis obliterans and rheumatoid arthritis, three of whom had received penicillamine therapy. After excluding all recognized causes of chronic airflow obstruction in 2,094 patients, Turton and co-workers detected ten patients with cryptogenic bronchiolitis obliterans, five of whom also had rheumatoid arthritis. Three of these had received penicillamine. In addition, there are reports of five rheumatoid patients in whom penicillamine has been strongly associated with the development of acute bronchiolitis, as well as some occasional cases in earlier studies. In lung function studies, almost all patients have shown profound airway obstruction, with highly diminished FEV1 and hyperinflation. The course of the disease has been fulminating in about one half of the cases described, and deaths due to bronchiolitis are not unusual. Further, a patient with a benign course was recently described by Herzog et al.

The symptoms of our patient were typical of bronchiolitis, as were the auscultatory findings. Marked hyperinflation was the prominent feature in her lung function studies, while the results from dry spirometer tests were completely normal. The course of her disease seems to be benign. In these respects she resembles a patient recently described by Herzog et al.

The intraluminal granulation polyps at the bronchiolar level seen in bronchiolitis of various etiologies are not seen in patients with rheumatoid arthritis. The granulomatous narrowing of bronchioles seen in our patient is very rare. Granulomas may reflect a cell-mediated immune response to antigen. Common granulomatous diseases of the lungs (tuberculosis, sarcoidosis, hypersensitivity pneumonias) can be excluded on clinical grounds in this case. Interestingly, granulomas have also been observed following injections of immune complexes.

The demonstration of IgM- and IgG-containing plasma cells in the bronchiolar walls of our patient suggests direct immune-mediated lung injury. Herzog et al reported in their patient with bronchiolitis and rheumatoid arthritis linear deposition of IgG in alveolar walls. Extensive IgM deposition in pulmonary arterioles, alveolar walls, and adjacent to the cavitary pulmonary rheumatoid nodules has been reported along with faint linear IgG alveolar wall staining in rheumatoid lungs.

Our patient had received penicillamine five years before her bronchiolitis was detected, so it may not have been the initial stimulus for the disease process. Her respiratory symptoms always developed two days after gold injections at the beginning of the pulmonary disease. When the resumed penicillamine therapy was stopped, her respiratory symptoms diminished and disappeared, and her lung function improved (Table 1). Probably patients with rheumatoid arthritis are more prone to develop bronchitis and bronchiolitis, and in such cases penicillamine may interfere with the usual healing process. The effect of gold in the development of bronchiolitis is completely unknown, although notably gold can cause nonspecific interstitial inflammation with fibrosis.

Although bronchiolitis in rheumatoid patients may be a rare condition, we advocate extreme care with rheumatoid patients receiving antirheumatic drugs who develop respiratory symptoms. Their lungs must be thoroughly examined, and the auscultatory findings themselves may be important for diagnosis. In our opinion, the measurement of lung volumes is the most sensitive lung function parameter in early bronchiolitis.

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Table 1—Results of Lung Function Studies

*Pathologic findings.

REFERENCES
7. Woitowitz HJ, Buchheim FW, Woitowitz R. Zur Theorie und Praxis der Ganzkörperplethysmographie in der Lungen-
Dissolution of Pulmonary Carcinoma via Argon-Laser Bronchoscopy

First Clinical Use of Fiberoptic Metal Cautery Cap Heated by Laser Radiation

Garrett Lee, M.D.; Richard Rubinson, M.D., F.C.C.P.; Ming C. Chan, M.D.; Daniel Stobbe; Robert L. Reis, M.D., F.C.C.P.; and Dean T. Mason, M.D., F.C.C.P.

A 60-year-old man with nonresectable lung cancer underwent bronchoscopy which revealed a large squamous cell tumor mass narrowing the free airway to the left lung. A flexible quartz fiber connected to an argon ion laser was then inserted through a hollow channel of the fiberoptic bronchoscope. The laser was activated to heat the metal cap on the distal tip of the fiber to thermally dissolve the mass and relieve airway obstruction.

Laser therapy has recently been utilized in clinical pulmonary oncology for relief of tracheobronchial stenosis. The available laser systems for such application have employed a bare-tipped fiber with recognized inherent hazards related to free-beam photodestruction. In our laboratories, we have previously devised a fiberoptic laser-heated metal cautery cap for dissolution of atherosclerotic lesions which also protects against vascular damage. The present report describes the extension of this new development of a metallic cap mounted at the distal end of a flexible quartz laser fiber for the controlled thermal vaporization of life-threatening nonresectable tumor producing airway obstruction.

Case Report

A 60-year-old white man who underwent right upper lobectomy one year previously for squamous cell lung carcinoma was admitted for recurrence of pulmonary cancer. His chief symptoms were shortness of breath and hemoptysis. The patient underwent bronchoscopy using topical tetracaine (Pontocaine) anesthesia. The proximal trachea was benign, but in the distal trachea, just cephalad to the carina, there was a large tumor mass obliterating the right mainstem bronchus and narrowing the free airway to the left lung.

To relieve the obstruction without thoracotomy, a flexible 400-µm central core quartz fiber with a metal cautery cap at its distal end was inserted into the flushing channel of a fiberoptic bronchoscope. The proximal end of the fiber was connected to an argon-ion laser source and using fiberoptic visual guidance, the distal tip was positioned in direct contact with the surface of the tumor. In Figure 1, the endobronchial tumor mass can be visualized along with the cautery cap protruding out the tip of the bronchoscope (top panel). To partially dissolve the mass, several bursts of 5-W laser energy were delivered to heat the cautery cap for 10 to 20 seconds. The middle and bottom panels of Figure 1 demonstrate the result of a single laser burst. Burned pieces of debrided tumor tissue adhered to the surface of the metal cap which were then removed (middle panel). In this way, the central portion of the tumor in the distal trachea was objectively cannalized allowing considerable enhancement of air passage to the left lung. The bronchoscope was withdrawn, and the patient tolerated the procedure extremely well without complications. Immediately upon completion of this new treatment modality, his dyspnea was abated concomitantly with the observed improvement of aeration to the left lung.

Discussion

The use of lasers in the treatment of endotracheal lesions offers rapid tissue vaporization with little bleeding, edema, and scar formation. However, present carbon dioxide laser systems usually necessitate general anesthesia and direct vision through a rigid bronchoscope. There are also the inherent dangers of retinal damage to the bronchoscopist and intratracheal fire hazards to the patient during the administration of oxygen and general anesthetics for laser surgery in the tracheobronchial tree. In addition, the administration of tumor-localizing hematoporphyrin to enhance argon laser photodestruction of endobronchial malignancies currently lacks the ability to penetrate deep into the tumor mass.

The application of a metal cautery cap connected to the distal tip of a flexible quartz fiber provides a number of advantages. It can be easily inserted through a flexible fiberoptic bronchoscope and positioned onto the tumor by fiberoptic visual guidance. The heated metal cap minimizes the risk of damaging surrounding normal tissue due to stray laser beams and eliminates the chance of retinal damage. Furthermore, it can be precisely directed and the vaporization process can occur deep into the tumor mass.

This represents the first report of the clinical use of a fiberoptic metal cautery cap heated by laser conduction. The original idea was based on our initial laboratory experiments in 1977 using an electrically heated metal cap device which was successful in dissolving atherosclerotic fatty-fibrous plaques. The vaporization of plaques by this improved metal...