scleroderma and pneumoconioses.5,7 The effect of smoking upon this membrane is less obvious, but the studies of Hubert et al4 with horseradish peroxidase indicate that the permeability of the epithelium of both the airways and alveoli rapidly increases in animals exposed to cigarette smoke and these changes are readily reversible. Clearances are also quite elevated in many patients with pulmonary parenchymal injury associated with adult respiratory distress syndrome, whereas these test results tend to be relatively normal in patients with pulmonary edema secondary to congestive heart failure.9 Rapid clearance is reported in animal models of oleic acid edema as compared to normal or elevated left atrial pressure.10 Neonatal respiratory distress syndrome also seems to be accompanied by increases in epithelial permeability.11

The widespread availability of both scanning equipment and 99mTc-radionuclides has made radioaerosol procedures accessible to most general hospitals. The introduction of new nebulizers which generate large numbers of very small droplets has simplified the manner in which these tests are conducted. The ease with which multiple views of the lung can be obtained with radioaerosols presents a distinct advantage over radioactive gases such as 133Xe in studies of regional ventilation.12,13 Measurement of radionuclide clearance involves little complexity other than determining the decline in radioactivity in the lungs for ten minutes.14 Rather than representing merely another test of gas transport, it yields new information relevant to the exchange of both solutes and fluid across the pulmonary epithelium. Among potential uses for the procedure which have yet to be investigated are the early detection of lung injury and the quantification of response to therapy. With the addition of alternative indicators and droplet size, it is likely that further applications for the procedure will be found over the coming years, but the clinical value of the approach already seems assured.

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Osler-Charcot Disease
A New Title For An Old Friend

The term Hamman-Rich syndrome refers to progressive interstitial fibrosis of the lungs of unknown cause. The original description includes four young patients who died of acute progressive dyspnea within six months.1 We know now that the disease is not always progressive nor fatal and that it is more common in the third and fourth decades of life. Purists immediately raised their banner and refused to use Hamman-Rich syndrome to describe chronic interstitial fibrosis.

Thus, subsequent to the description of the disease by Hamman and Rich, various terms have sprung up like mushrooms in the night. These include: cirrhosis of the lung, chronic interstitial pneumonia, chronic interstitial pneumonitis, fibroid pneumonia, fibro-cystic pulmonary dysplasia, idiopathic interstitial pneumonitis and idiopathic pulmonary fibrosis. Scadding, in order to clear this semantic confusion, proposed that the condition be called fibrosing alveolitis because the inflammatory and fibrotic changes predominantly affect alveolar walls, as opposed to alveolar spaces.2 Scadding3 argued that the word interstitial should be dropped and cryptogenic added, reflecting...
our ignorance of the cause. Cryptogenic fibrosing alveolitis, thus, became an accepted term in Europe and the British Isles. However, because the term interstitial pulmonary fibrosis is so entrenched, it is unlikely to be totally superseded by fibrosing alveolitis, even though this may be more accurate.

What then are we to do? Purists will not accept the term Hamman-Rich syndrome. Our colleagues across the Atlantic would prefer cryptogenic fibrosing alveolitis. However, on the North American continent, idiopathic pulmonary fibrosis continues to remain popular.

Here is an observation worth pondering. While going through William Osler’s textbook of medicine, I came across the following paragraph:

In one of Charcot’s cases . . . death occurred about three months and a half after the onset of the acute disease and the lung was two thirds of the normal size, grayish in color, and hard as cartilage. In the only case of the kind which has come under my observation the patient died about a month from the onset of the chill. The lung was uniformly solid and grayish in color. Microscopically these areas showed advanced fibroid changes and great thickening of the alveolar cells . . .

This, indeed, is a perfect description of interstitial pulmonary fibrosis by Osler, more than 50 years before the publication of Hamman and Rich, from the John Hopkins Hospital. By calling idiopathic pulmonary fibrosis (cryptogenic fibrosing alveolitis) Osler-Charcot disease, we not only will be clearing up the semantic confusion, but also will be awarding credit to those who observed and described this entity first. I suggest that the entity, which has roamed so long in search of a title, be now duly crowned as Osler-Charcot disease.

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William Tell and Technology*

A 14th century legend recounts that William Tell was forced to shoot an apple from atop his son’s head when he refused to pay homage to the governor. A man of great skill with nerves of steel, Tell was very much like our modern bronchoscopists. Armed with 20th century lances and as much concern for their patients as Tell had for his son, they also challenge the status quo. In this issue (see page 573) Schwartz et al describe the piercing of a bronchogenic cyst using sophisticated radiologic targeting to guide an endoscopically directed needle.

Bronchogenic cysts, like apples, are round structures usually found in the periphery of the tracheobronchial tree. About one-third are found near the major branches in the mediastinum. They may even fall off the tree and land in the pericardium, esophagus, or the vertebral column. In infancy and childhood, these cysts commonly present with symptoms or signs of respiratory distress, dyspnea, or chest discomfort due to the mass effect. In adults, they are often asymptomatic, although they may present with infection, hemorrhage, perforation and, rarely, malignant degeneration.

Prior to the development of computerized axial tomography (CAT scanning) and the transbronchial needle, these lesions were suspect on conventional tomography because of their spherical shape, homogeneous density and position in the chest. The absence of more refined radiographic and endoscopic techniques led to the performance of major surgical procedures to obtain a diagnosis and occasionally to relieve symptoms. Combining the computerized axial tomogram with the transbronchial needle aspiration technique appears to be an ideal solution to the diagnostic dilemma posed by these congenital lesions. The low density readings often, but not always, found with CAT scanning raises our level of suspicion that we are seeing a bronchogenic cyst. Because these lesions seldom are malignant, it is probably unnecessary that they be removed unless symptoms are present or cytologic aspirates show malignant cells.

The uses of the transbronchial needle aspiration technique are expanding, as evidenced by the report of Schwartz et al. In the future, the transbronchial needle aspiration technique may serve not only our diagnostic needs, but may also be therapeutic. If a large enough volume of fluid can be removed from a bronchogenic cyst via needle aspiration, then symptoms related to the mass effect may disappear and surgery may be avoided. We must await reports of the feasibility of using the needle aspiration technique in the pediatric population.

The use of these new techniques requires the skill of a William Tell and intimate knowledge of the situation in which it is used. The analogy with William Tell must end here, for Tell promised that if he failed with his first arrow, the second would pierce the heart (of the governor).

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