The recognition that chronic thromboembolic disease is a treatable cause of pulmonary hypertension is relatively recent. Twenty-five years ago, only the University of California, San Diego, had an active program of pulmonary thromboendarterectomy. A common comment was that the disease was extremely rare, that it did not even exist in some countries, and that surgery for the condition was dangerous and rarely successful.

Times have changed. With the increased recognition of both the occurrence of the condition and the efficacy of treatment, many centers, both in the United States and internationally, have active programs, and an international registry has been created to assess operability and results.

The occluding material in chronic thromboembolic disease is fibrotic scarlike scar tissue, the result of organizing clot (Figure 1). It may be partially or totally obstructive, but more than 50% of the pulmonary vasculature must be affected for pulmonary hypertension to occur. Treatment is by surgical removal (pulmonary thromboendarterectomy), balloon angioplasty, pulmonary vasodilating agents, or some combination. Only surgical endarterectomy offers the possibility of cure. Balloon angioplasty and vasodilators are helpful but rarely curative, because balloon angioplasty cannot open a totally occluded vessel with no runoff, which is often the case.

It has long been surmised that thromboembolism and thromboembolic disease have a different incidence and course in Japan to that usually seen in Europe and the United States. The article in this issue of the Journal by Chausheva and colleagues attempts to address this issue. In Austria, there is a high awareness of the incidence, diagnosis, and treatment of chronic thromboembolic disease, whereas this awareness is not as acute (yet) in Japan. This underlying difference may explain some of the stated differences in the incidence of the disease and the selection of patients for operation. The technique of balloon angioplasty was pioneered in Japan and in part grew out of the small experience in operative treatment in that country.

Even in Europe, experience in pulmonary thromboendarterectomy is still growing. In the registry, fewer than two-thirds of patients with the condition diagnosed underwent surgery. In the United States, and particularly at the University of California, San Diego, it is extremely rare for a patient to be denied surgery if the condition is diagnosed. In our experience of now more than 4000 operated cases done in the course of 30 years by 4 surgeons, we have found that experience in several hundred cases is required for the confidence and ability to address very distal (subsegmental)
occlusions with surgery. These are lesions that in many other centers are declared “inoperable” or only amenable to balloon angioplasty or drug therapy. It is likely that, as surgical experience in other centers grows, the number of patients in the registry with disease that is declared “inoperable” will fall.

Although the authors are well respected in the field, the study of Chausheva and colleagues\(^3\) suffers from the relatively small number of patients being studied. The pathologic characteristics of the resected specimens have been carefully assessed, but it is still difficult to be sure that the type and incidence of chronic thrombotic occlusion of the pulmonary vasculature are different between Austria and Japan. Time will tell.

References