Operative Techniques in
Thoracic and
Cardiovascular Surgery

Pulmonary Endarterectomy for Chronic Thromboembolic Disease

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Pulmonary thromboendarterectomy (PTE) is the definitive treatment for chronic pulmonary hypertension as the result of thromboembolic disease. Although pulmonary embolism is one of the more common cardiovascular diseases affecting Americans, PTE remains an uncommon procedure, primarily because this form of chronic pulmonary hypertension remains an underdiagnosed condition. Recently there has been an increasing awareness of recognizing chronic pulmonary thromboembolic disease as a cause of severe pulmonary hypertension. Although the disease is estimated to occur in approximately 1 to 5% of all patients who have previously developed an acute pulmonary embolism, the true prevalence is suspected to be much higher. Chronic thromboembolic pulmonary hypertension (CTEPH) is characterized by intraluminal thrombus organization, fibrotic scar-tissue-like stenosis, and the ensuing vascular remodeling of unaffected pulmonary vessels. Pulmonary endarterectomy is an operation that is considered curative for thromboembolic pulmonary hypertension. The only other alternative surgical procedure for treatment of pulmonary hypertension is transplantation. Considering the curative nature of PTE, long-term affects of transplantation, its strict medical regimen and undesirable side effects, as well as scarce donor availability, it is clear that PTE is a superior option to transplantation for this condition.

Patients affected by CTEPH may present with a variety of debilitating cardiopulmonary symptoms. Considerable progress has been made over the past decade in understanding the etiology, prevalence, natural history, and therapeutic approach to chronic thromboembolic pulmonary hypertension. Acute pulmonary thromboembolism and its chronic sequelae are significant causes of morbidity and mortality in the United States and the world. However, the chronic disease process, even when established and symptomatic, is notoriously underdiagnosed because of the nonspecific nature of the two major symptoms, effort dyspnea and fatigue, and the fact that physical findings may be elusive until right heart failure occurs. Calculations extrapolated from mortality rates and the random incidence of major thrombotic occlusion of pulmonary vessels at autopsy support an estimate that more than 100,000 people in the United States currently suffer from pulmonary hypertension that could be relieved by operation.[1]

Once chronic pulmonary hypertension develops, the prognosis is poor, and it is worse for those who do not have intracardiac shunts. Thus, patients with primary pulmonary hypertension and those with pulmonary hypertension due to pulmonary emboli fall into a higher risk category than those with Eisenmenger's syndrome and encounter a higher mortality rate. In fact, once the mean pulmonary pressure in patients with thromboembolic disease reaches 50 mm Hg or more, the 3-year mortality approaches 90%.[2] Therefore, despite an improved understanding of pathogenesis, diagnosis, and management, pulmonary emboli and the long-term sequelae of thromboembolic pulmonary hypertension remain frequent and often fatal disorders.

Pulmonary endarterectomy is a technically demanding operation currently performed in only a few select centers around the world. Proper patient selection, meticulous surgical technique, and careful postoperative management have contributed to the effectiveness of this operation. A true endarterectomy (not an embolectomy) of all affected segments of the lung is essential to clear all affected areas of the pulmonary vasculature. The procedure, when performed at an experienced center, ameliorates pulmonary hypertension by improving lung ventilation–perfusion mismatch, significantly improving right ventricular dysfunction and tricuspid regurgitation, limiting retrograde expansion of thromboembolic material, and preventing arteriopathic changes in the remaining patent small pulmonary vessels.

In this article, we describe the surgical procedure as it is performed at the University of California San Diego (UCSD) Medical Center. To date, the largest surgical volume of pulmonary endarterectomy has been performed at our center, and it is this experience that forms the basis of this report.

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Operative Technique

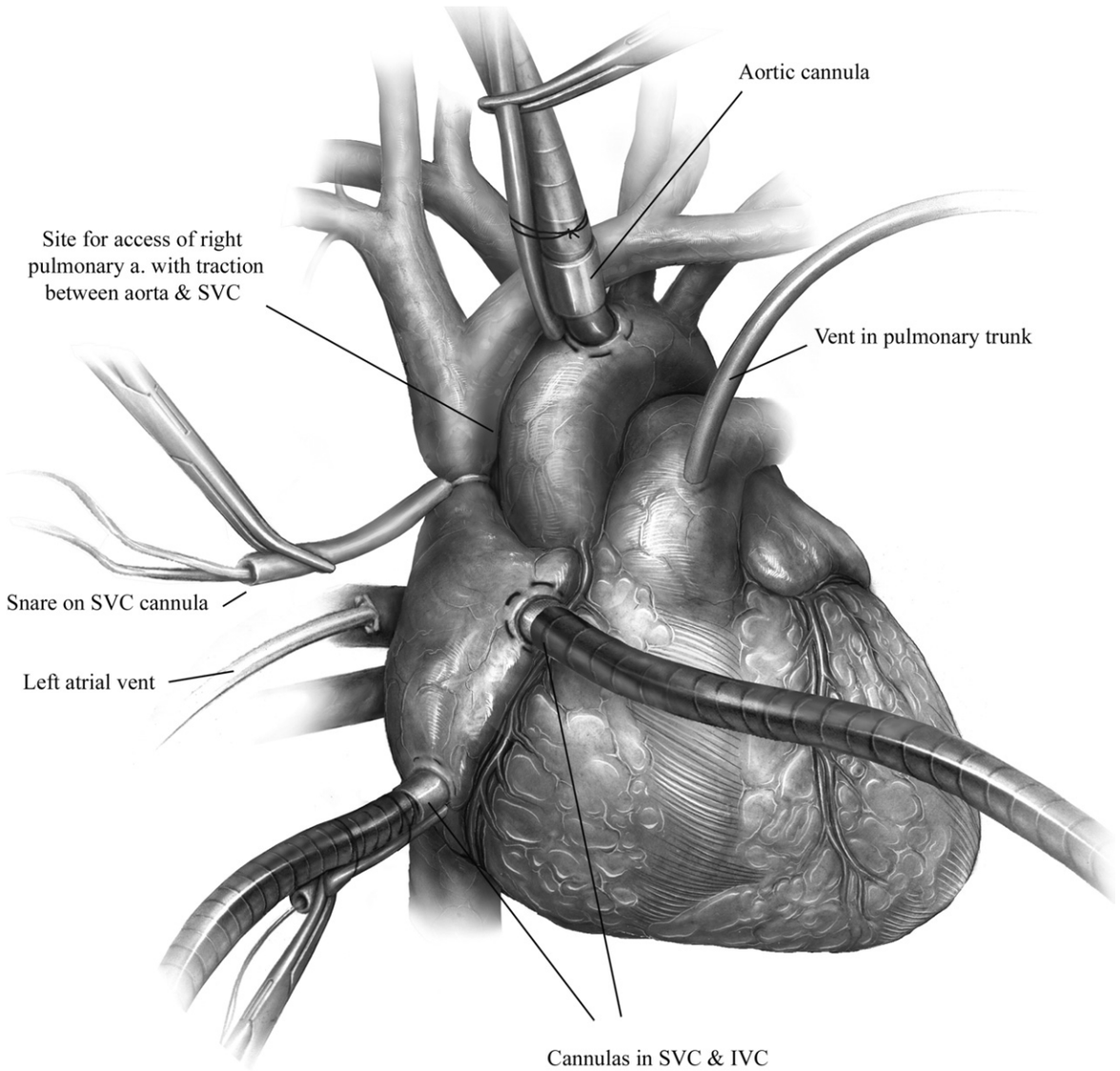

Figure 1 After a median sternotomy is made, the pericardium is incised longitudinally and attached to the wound edges. Full cardiopulmonary bypass is instituted with high ascending aortic cannulation and two caval cannulae. The heart is emptied on bypass, and a temporary pulmonary artery vent is placed in the midline of the main pulmonary artery about 1 cm distal to the pulmonary valve. Patient is then cooled to 20°C with the pump oxygenator; surface cooling with both the head jacket and the cooling blanket is also begun. When ventricular fibrillation occurs, an additional vent is placed in the left atrium through the right superior pulmonary vein. Initially, it is most convenient for the primary surgeon to stand on the patient's left side and perform the endarterectomy on the right side. IVC = inferior vena cava; SVC = superior vena cava; a = artery.

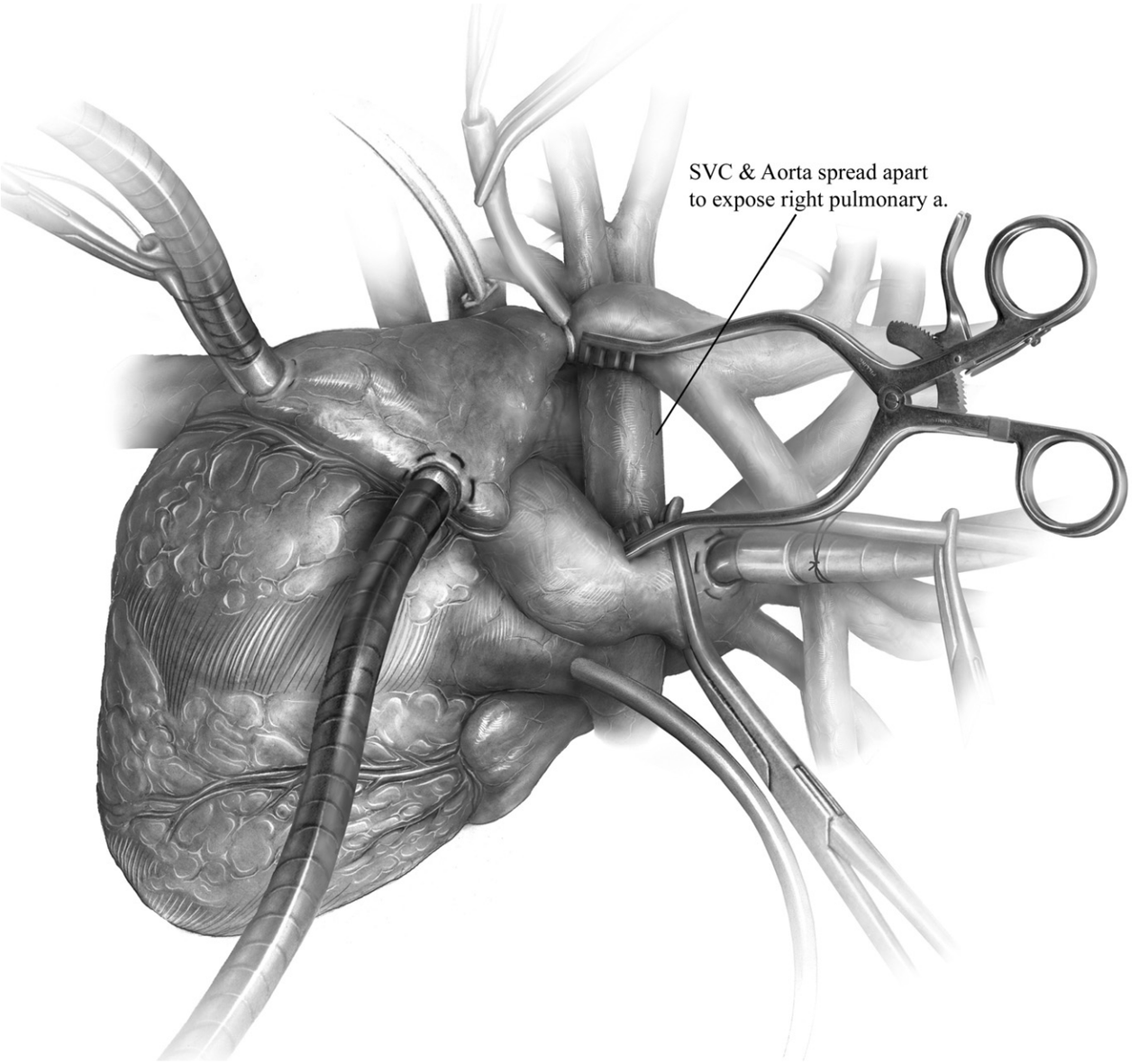

Figure 2 The approach to the right pulmonary artery is made medial, not lateral, to the superior vena cava. During the cooling period, some preliminary dissection can be performed, with full mobilization of the right pulmonary artery from the ascending aorta. The superior vena cava is also fully mobilized. Once the core temperature has reached 20°C, an aortic cross-clamp is applied and myocardial protection is provided through a single dose of antegrade cold blood cardioplegia (1 L). The entire procedure is now performed with a single aortic cross-clamp period with no further administration of cardioplegic solution. Additional myocardial protection is provided by using a cooling jacket surrounding the heart throughout the remainder of the procedure. Both tourniquets are now secured around the superior and inferior venae cavae to ensure complete drainage and to avoid any air entry in the venous cannulae during circulatory arrest. SVC = superior vena cava; a = artery.

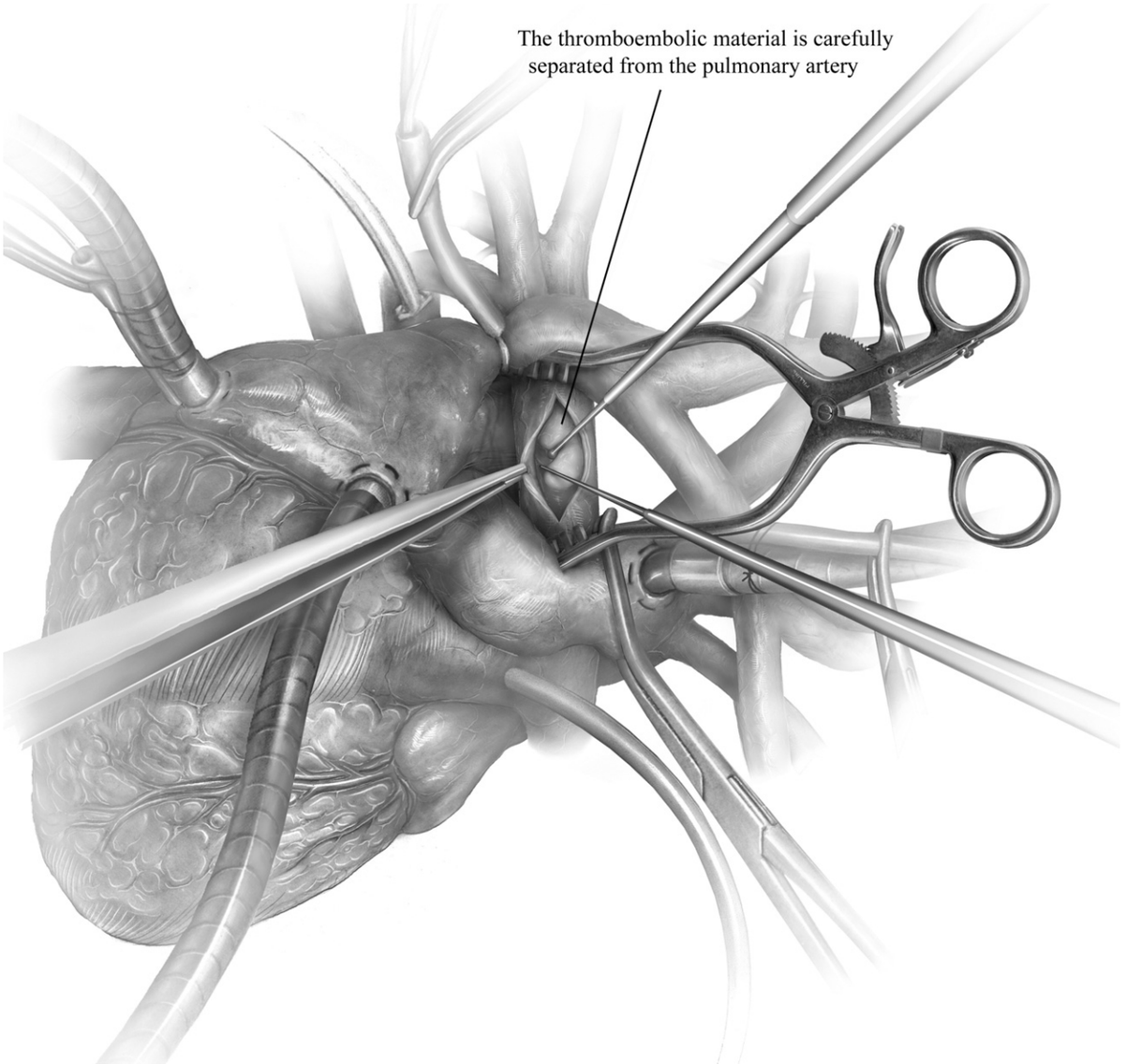

Figure 3 A modified cerebellar retractor is then used to expose the pulmonary artery between the aorta and the superior vena cava. An incision is made in the right pulmonary artery from beneath the ascending aorta out under the superior vena cava and entering the lower lobe branch of the pulmonary artery just after the takeoff of the middle lobe artery. It is important that the incision stays in the center of the vessel and continues in the middle of the descending pulmonary artery into the lower.

Any loose thrombus, if present, is now removed. This is necessary to obtain good visualization. It is most important to recognize, however, that first, an embolectomy without subsequent endarterectomy is quite ineffective and, second, that, in most patients with chronic thromboembolic hypertension, direct examination of the pulmonary vascular bed at operation generally shows no obvious embolic material. If the bronchial circulation is not excessive, the endarterectomy plane can be found during this early dissection. However, although a small amount of dissection can be performed before the initiation of circulatory arrest, it is unwise to proceed unless perfect visibility is obtained because the development of a correct plane is essential.

The correct plane appears pearly white, which is smooth and silky in appearance and lies between the intima and media. A microtome knife is used to develop the endarterectomy plane posteriorly, because any inadvertent egress in this site could be repaired readily, or simply left alone. Once the plane is correctly developed, the specimen is trimmed as to leave an area of full thickness next to the incision line for subsequent repair and closure.

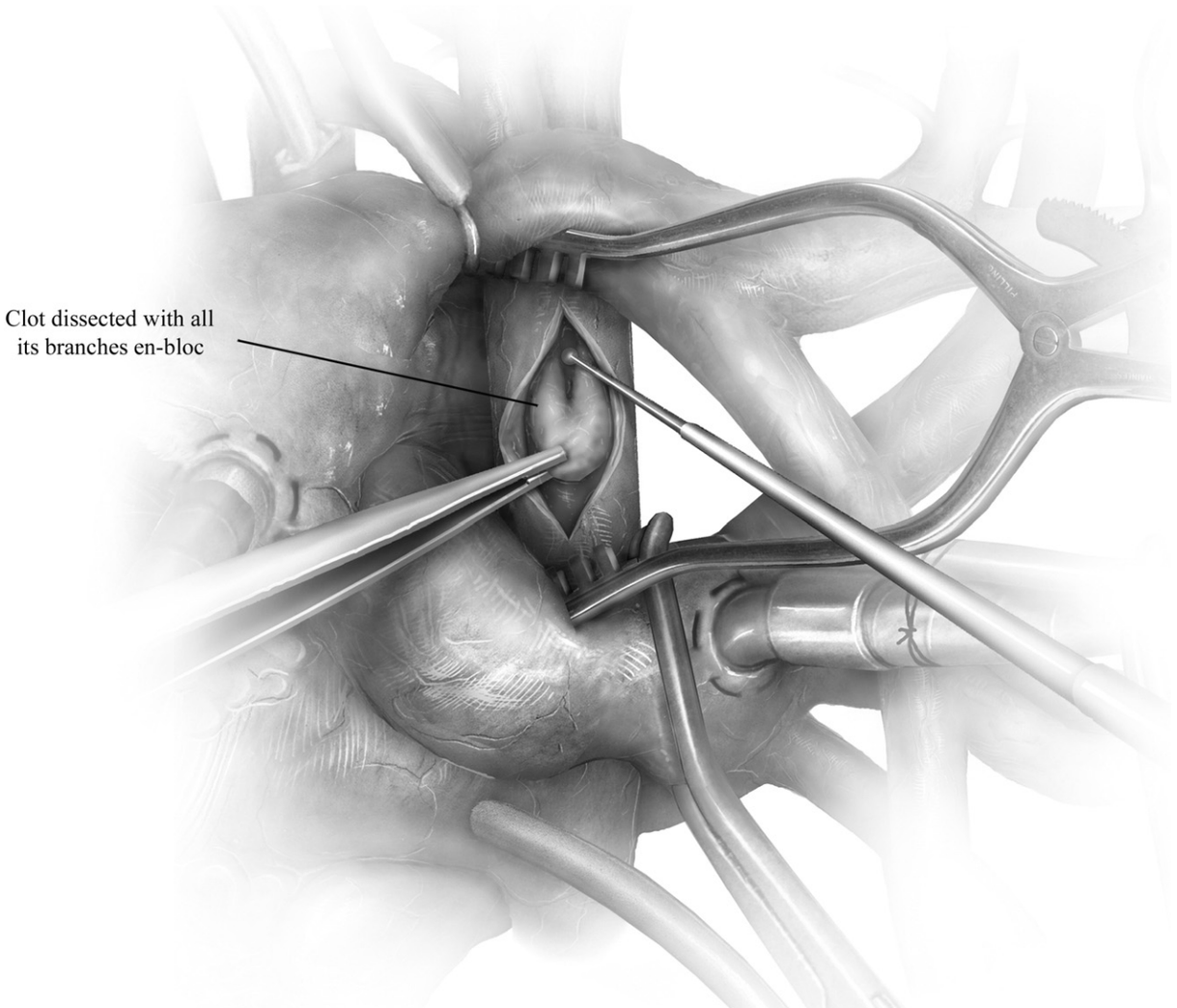

Figure 4 With the modified cerebellar retractor in place and the artery well exposed, the dissection is then carried out. When blood obscures direct vision of the pulmonary vascular bed, thiopental is administered (500 mg to 1 g) until the electroencephalogram becomes isoelectric. Circulatory arrest is then initiated, and the patient undergoes exsanguination. All monitoring lines to the patient are turned off to prevent the aspiration of air. Snares are tightened around the cannulae in the superior and inferior venae cavae. It is rare that one 20-minute period for each side is exceeded.

The endarterectomy is then performed with an eversion technique. Because the vessel is everted and subsegmental branches are being worked on, a perforation here will become completely inaccessible and invisible later. The specimen depicted in this figure is grossly enhanced to demonstrate the surgical technique; in fact, most thromboembolic specimens are much finer and more distal in the segmental and subsegmental branches. This is why the absolute visualization in a completely bloodless field provided by circulatory arrest is essential. It is important that each subsegmental branch is followed and freed individually until it ends in a "tail," beyond which there is no further obstruction. Residual material should never be cut free; the entire specimen should "tail off" and come free spontaneously.

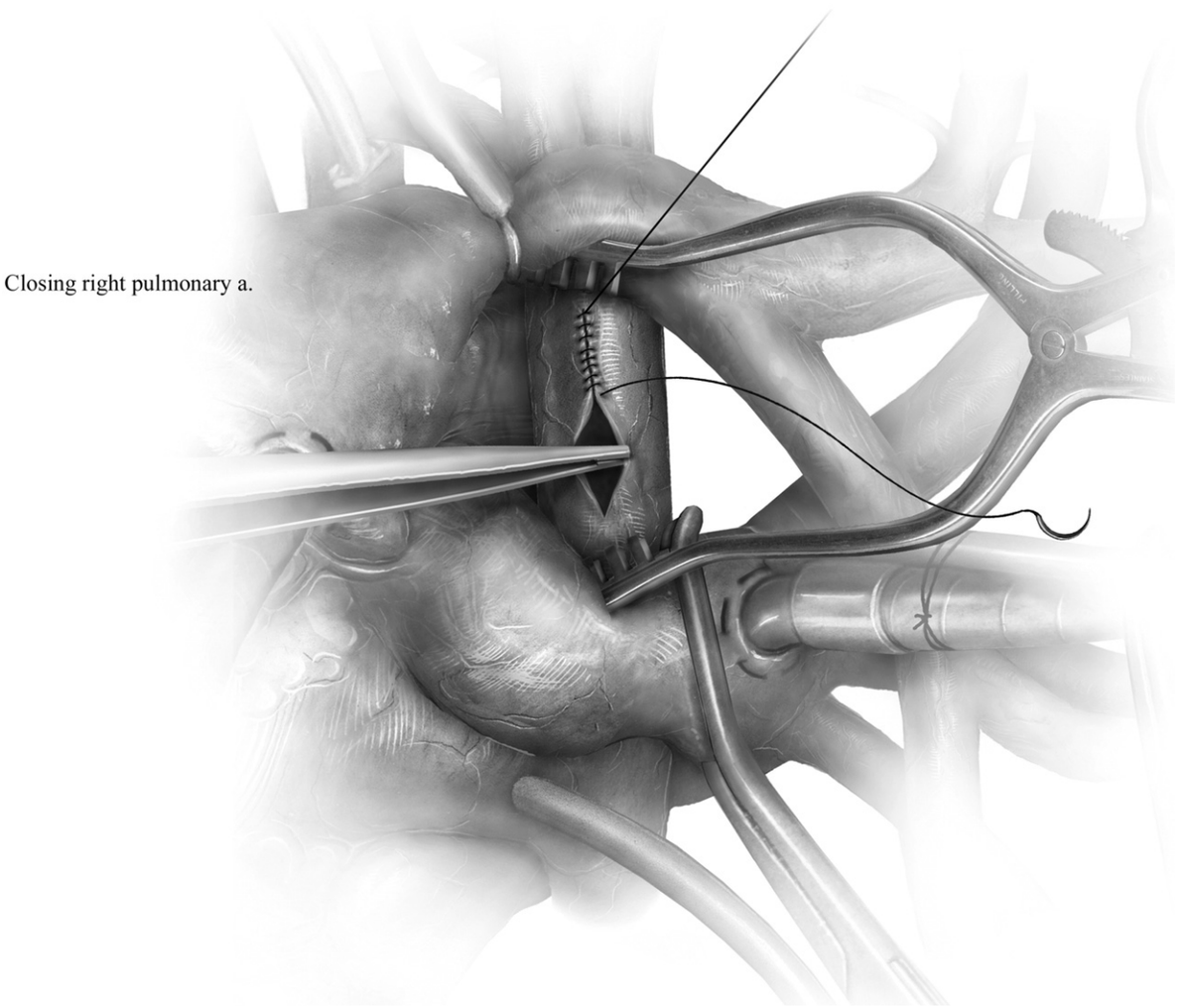

Figure 5 Once the right-sided endarterectomy is completed, circulation is restarted, and the arteriotomy is repaired with a continuous 6-0 polypropylene suture. The hemostatic nature of this closure is aided by the nature of the initial dissection, with the full thickness of the pulmonary artery being preserved immediately adjacent to the incision. a = artery.

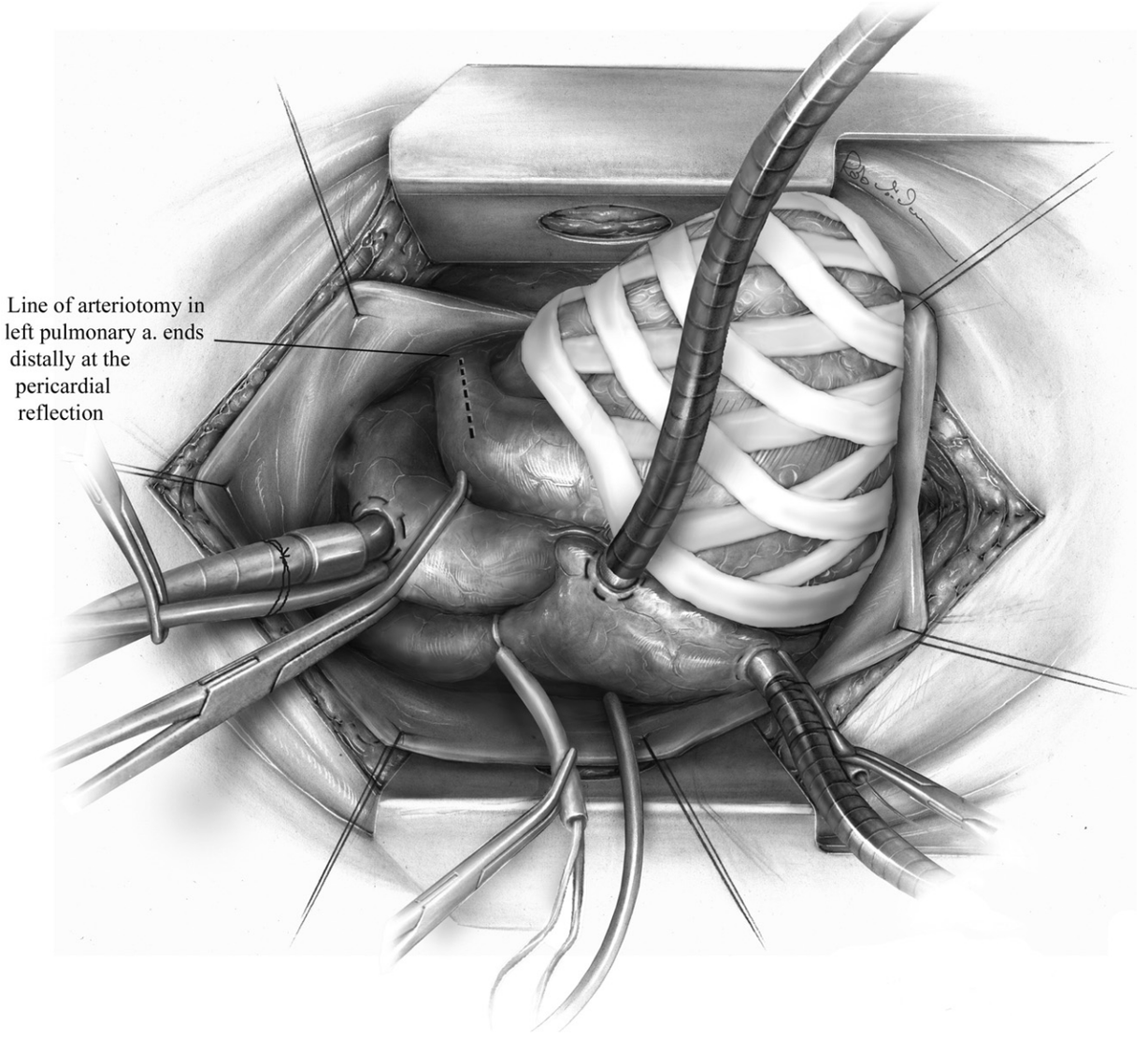

Figure 6 After the completion of the repair of the right arteriotomy, the surgeon moves to the patient's right side. The pulmonary vent catheter is withdrawn, and an arteriotomy is made from the site of the pulmonary vent hole laterally to the pericardial reflection, avoiding entry into the left pleural space. Neither pleural cavity is entered. Care must be taken to avoid injury to the left phrenic nerve. Additional lateral dissection does not enhance intraluminal visibility, may endanger the left phrenic nerve, and makes subsequent repair of the left pulmonary artery more difficult. The heart is wrapped within a cooling jacket and retracted using a mesh-like basket retractor shown in the figure. a = artery.

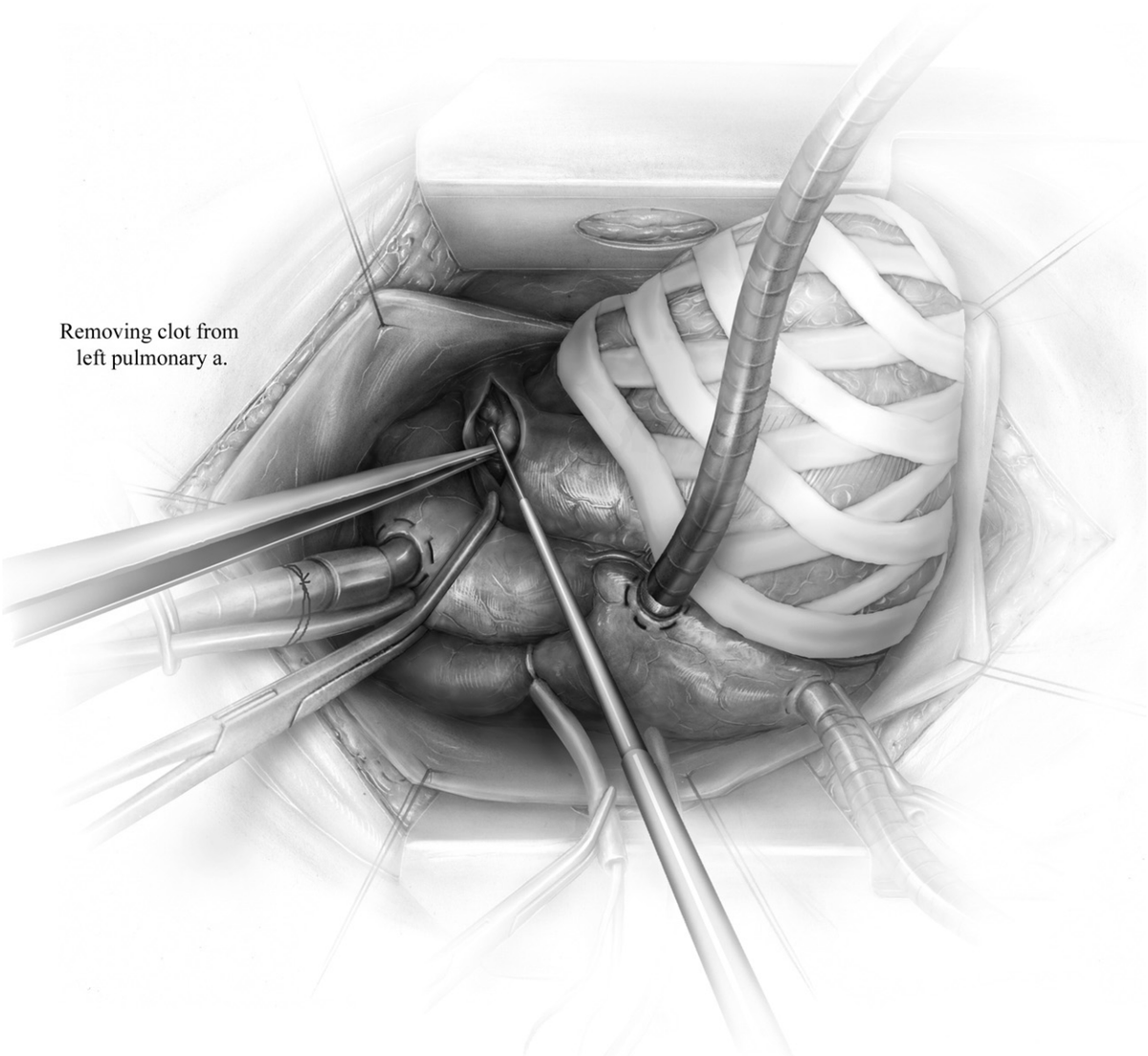

Figure 7 The left-sided dissection is virtually analogous in all respects to that accomplished on the right. The main obstructing material is a thickened scar-like tissue that obstructs each branch distally, and removal of only gross thrombus visible on initial inspection will be ineffective. The plane of dissection is developed posteriorly, and again the specimen is trimmed around the edges of the incision for subsequent closure. The duration of circulatory arrest intervals during the performance of the left-sided dissection is subject to the same restriction as the right. The specimen is followed in each segmental and subsegmental branch to ensure complete removal of the endarterectomy material. Whenever possible, the left lower lobe should be dissected circumferentially and the specimen followed distally. The left main bronchus crosses anteriorly and it is extremely important to have firm grasp of the specimen at this location. If the specimen breaks, it is particularly difficult to regain exposure of these distal branches. a = artery.

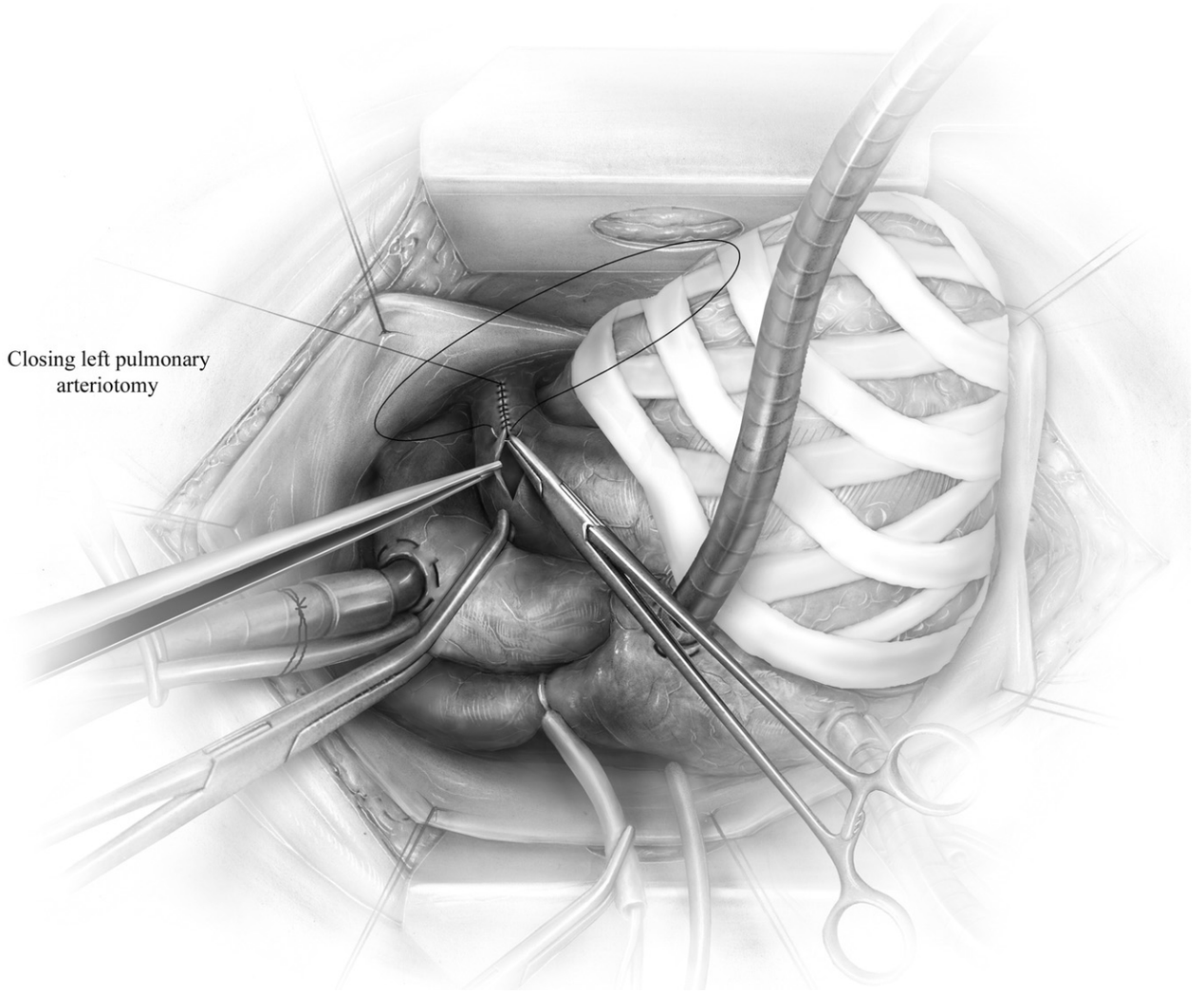

Figure 8 After completion of the endarterectomy, cardiopulmonary bypass is reinstituted and warming is commenced. During warming, a 10°C temperature gradient is maintained between the perfusate and body temperature. The rewarming period generally takes approximately 90 to 120 minutes but varies according to the body mass of the patient. When the left pulmonary arteriotomy has been repaired, the pulmonary artery vent is replaced at the top of the incision. The heart is retracted upwards and to the left, and a posterior pericardial window is made, between the aorta and the left phrenic nerve. Alternatively a soft drain can be left behind the heart to drain any developing posterior collection.

If other cardiac procedures are required, such as coronary artery or mitral or aortic valve surgery, these are conveniently performed during the systemic rewarming period. The most common concomitant procedure is closure of foramen ovale, followed by coronary bypass surgery. Although tricuspid valve regurgitation is invariable in these patients and is often severe, tricuspid valve repair is not performed. Right ventricular remodeling occurs within a few days, with the return of tricuspid competence. At the completion of all associated procedures, myocardial cooling is discontinued. The left atrial vent is removed, and the vent site is repaired. All air is removed from the heart, and the aortic cross-clamp is removed.

When the patient has fully rewarmed, cardiopulmonary bypass is discontinued. With a successful endarterectomy, the cardiac output is generally high, with a low systemic vascular resistance. Temporary atrial and ventricular epicardial pacing wires are placed. Wound closure is routine and similar to other cardiac procedures.

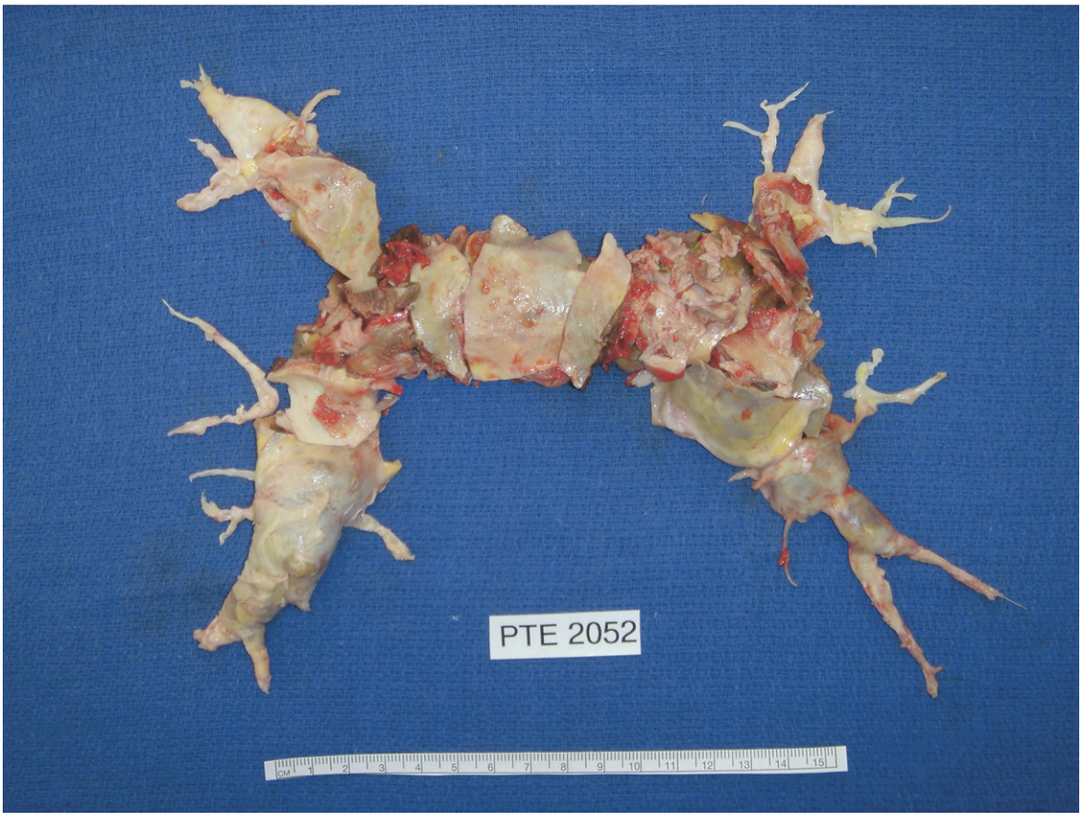

Figure 9 Specimen removed from both lungs during pulmonary endarterectomy. The ruler measures 15 cm. Note that removal of proximal thromboembolic material without endarterectomy of all the distal branches would leave significant disease behind, without any relief in pulmonary pressures or the pulmonary vascular resistance. The above specimen is characterized as Type I disease. There are four broad types of pulmonary occlusive disease related to thrombus that can be appreciated. Type I disease (approximately 20% of cases) refers to the situation in which major vessel clot is present and readily visible on the opening of the pulmonary arteries. In Type II disease (approximately 60% of cases), no major vessel thrombus can be appreciated. Type III disease (approximately 20% of cases) presents the most challenging surgical situation. The disease is very distal and confined to the segmental and subsegmental branches. No occlusion of vessels can be seen initially. The endarterectomy plane must be carefully and painstakingly raised in each segmental and subsegmental branch. Type IV disease (see Comments) does not represent primary thromboembolic pulmonary hypertension and is inoperable. In this entity, there is intrinsic small-vessel disease, although secondary thrombus may occur as a result of stasis. (Color version of figure is available online at http://www.us.elsevierhealth.com/optechstcvs.)

Comments

Pulmonary hypertension caused by chronic pulmonary embolism is underrecognized and carries a poor prognosis. Medical therapy for this condition is palliative at best and only transiently improves symptoms. Because of the obstructive nature of this disease, the only curative option is to remove all the obstructive thromboembolic material, by means of pulmonary endarterectomy. The only therapeutic alternative to pulmonary endarterectomy is lung transplantation; however, we consider this inappropriate use of resources and an inferior option. The advantages of pulmonary endarterectomy include a lower operative mortality (3-month survival for lung transplantation for pulmonary hypertension 1990 to 2001, 72%; for heart–lung transplantation for pulmonary hypertension, 70%[3,4]), better long-term survival, and the avoidance of chronic immunosuppressive treatment and allograft rejection. Currently our survival rate for pulmonary endarterectomy at UCSD is 95.7% (2005 to 2006), and the operation can provide sustained clinical benefit. Considering these results, clearly pulmonary endarterectomy is the procedure of choice and much more advantageous than lung transplantation for curative treatment of this disease. Although through our efforts more and more centers are recognizing this procedure as the treatment of choice and are avoiding inappropriate lung transplantation, the main problem remains that the condition itself continues to go unrecognized. The patients are generally misdiagnosed and mistreated for many years before they are truly identified as suffering from chronic thromboembolic pulmonary hypertension.

It is noteworthy to point out that, despite favorable results in our series of over 2000 pulmonary endarterectomies, we have come to realize that a small subset of patients do not benefit from this operation, as they manifest solely arteriolar-capillary vasculopathy similar to idiopathic pulmonary arterial hypertension. In general these patients do not have any thromboembolic disease and in essence were actually misdiagnosed; however, they are hard to distinguish from their counterpart. The main problem is the distal vasculopathy, and as such, they do not benefit from pulmonary endarterectomy. With our growing experience, we are accepting higher risk patients and some with minimal disease. Our philosophy is that as long as there is any evidence of thromboembolic material on preoperative studies, the patients are accepted for surgery if there are no other contraindications. The risk factor that characterizes patients with vasculopathy is high pulmonary pressures and pulmonary vascular resistance (PVR) without much evidence of thromboembolic obstruction on angiogram. Unresolved PVR is a significant risk factor for postoperative morbidity and mortality. In fact, the mortality rate is about 25% when the residual PVR after operation remains higher than 500 dyn s cm^{-5}, but only 2.7% when it was below this level. Further refinements in preoperative work-up, including pulmonary waveform analysis, pulmonary angiography, commuted tomography scanning, and magnetic resonance imaging, will help to further distinguish these patients from those truly affected by obstructive disease.[5-7]

Although pulmonary endarterectomy is technically demanding for the surgeon, mainly because it requires careful dissection of the pulmonary artery wall planes and the use of circulatory arrest; excellent results can be achieved. Improvements in operative technique developed over the last three decades allow pulmonary endarterectomy to be offered to patients with low mortality rate and anticipation of clinical improvement. Currently, at our institution, no patient is denied operation based on severity of pulmonary hypertension or age, as long as there is evidence of thromboembolic disease. With the earlier and more accurate diagnosis of patients presenting with thromboembolic pulmonary hypertension and more common knowledge that pulmonary endarterectomy is a safe and quite an effective operation for this condition, we anticipate that the utility of this procedure will only increase in the future.

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