An Insider's Guide to Pulmonary Thromboendarterectomy: Proven Techniques to Achieve Optimal Results

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Pulmonary thromboendarterectomy is the definitive treatment for chronic pulmonary hypertension as the result of thromboembolic disease. Although pulmonary embolism (PE) is one of the more common cardiovascular diseases affecting Americans, pulmonary thromboendarterectomy remains an uncommon procedure, mainly because this form of chronic pulmonary hypertension remains an underdiagnosed condition. These patients may present with a variety of debilitating cardiopulmonary symptoms. However, once diagnosed there is no curative role for medical management, and surgery remains the only option.

The exact incidence of PE remains unknown, but there are some valid estimates. Acute PE is the third most common cause of death (after heart disease and cancer). Approximately 75% of autopsy-proven PE is not detected clinically.[1] Dalen and Alpert[2] calculated that PE results in 630,000 symptomatic episodes in the United States yearly, making it about half as common as acute myocardial infarction, and three times as common as cerebral vascular accidents. This is, however, a low estimate, since in 70% to 80% of the patients where the primary cause of death was PE, premortem diagnosis was unsuspected.[3,4] The disease is particularly common in hospitalized elderly patients. Of hospitalized patients who develop PE, 12% to 21% die in the hospital, and another 24% to 39% die within 12 months.[5-7] Thus approximately 36% to 60% of patients who survive the initial episode live beyond 12 months, and may present later in life with a wide variety of symptoms. More than 90% of clinically detected pulmonary emboli are associated with lower extremity deep vein thrombosis (DVT), but in two-thirds of patients with DVT and PE, the DVT is asymptomatic.[8,9] Greenfield[10] estimates that approximately 2.5 million Americans develop DVT each year.

The prognosis for patients with pulmonary hypertension 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 mmHg or more, the 3-year mortality approaches 90%.[11] Surgical options are dependent on both the primary disease process and the reversibility of the pulmonary hypertension. With the exception of thromboembolic pulmonary hypertension, lung transplantation is the only effective therapy for patients with pulmonary hypertension, when the disease reaches end stage. Pulmonary transplantation is also still used in some centers as the treatment of choice for those with thromboembolic disease. However, a true assessment of the effectiveness of any therapy should take into account the total mortality once the patient has been accepted and put on the waiting list. Thus, the mortality for transplantation (and especially double-lung or heart-lung transplantation) as a therapeutic strategy is much higher than is generally appreciated because of the significant loss of patients awaiting donors. Considering the long-term use of antirejection medications with their associated side effects, the higher operative morbidity and mortality, the long waiting time, and inferior prognosis even after transplantation, transplantation is clearly an inferior option to pulmonary thromboendarterectomy. We consider it to be inappropriate therapy for this disease.

Pulmonary Thromboendarterectomy: Indications

Although there were previous attempts, Allison et al[12] did the first successful pulmonary "thromboendarterectomy" through a sternotomy using surface hypothermia, but only fresh clots were removed. The operation was done 12 days after a thigh injury that led to PE, and there was no endarterectomy. Since then, there have been many occasional surgical reports of the surgical treatment of chronic pulmonary thromboembolism,[13, 14] but most of the surgical experience in pulmonary endarterectomy has been reported from the UCSD Medical Center. Braunwald commenced the UCSD experience with this operation in 1970, which now totals more than 1500 cases. The operation described below[15], using deep hypothermia and circulatory arrest, is now the standard procedure.

When the diagnosis of thromboembolic pulmonary hypertension has been firmly established, the decision for operation is made based on the severity of symptoms and the general condition of the patient. Early in the pulmonary endarterectomy experience, Moser and colleagues[16] pointed out that there were three major reasons for considering thromboendarterectomy: hemodynamic, alveolo-respiratory, and prophylactic. The hemo-

dynamic goal is to prevent or ameliorate right ventricular compromise caused by pulmonary hypertension. The respiratory objective is to improve respiratory function by the removal of a large ventilated but unperfused physiologic dead space. The prophylactic goal is to prevent progressive right ventricular dysfunction or retrograde extension of the obstruction, which might result in further cardiorespiratory deterioration or death.[16] Our subsequent experience has added another prophylactic goal: the prevention of secondary arteriopathic changes in the remaining patent vessels. Most patients who undergo operation are within New York Heart Association (NYHA) class III or class IV. The ages of the patients in our series have ranged from 8 to 85 years. A typical patient will have a severely elevated pulmonary vascular resistance (PVR) level at rest, the absence of significant comorbid disease unrelated to right heart failure, and the appearance of chronic thrombi on angiography that appear to be in balance with the measured PVR level. Exceptions to this general rule, of course, occur.

Although most patients have a PVR level in the range of 800 dynes/sec/cm-^{5} and pulmonary artery pressures less than systemic, the hypertrophy of the right ventricle that occurs over time makes pulmonary hypertension to suprasystemic levels possible. Therefore, many patients (perhaps 20% in our practice) have a level of PVR in excess of 1000 dynes/sec/cm-^{5} and suprasystemic pulmonary artery pressures. There is no upper limit of PVR level, pulmonary artery pressure, or degree of right ventricular dysfunction that excludes patients from operation. We have become increasingly aware of the changes that can occur in the remaining patent (unaffected by clot) pulmonary vascular bed subjected to the higher pressures and flow that result from obstruction in other areas. Therefore, with the increasing experience and safety of the operation, we are tending to offer surgery to symptomatic patients whenever the angiogram demonstrates thromboembolic disease. A rare patient might have a PVR level that is normal at rest, although elevated with minimal exercise. This is usually a young patient with total unilateral pulmonary artery occlusion and unacceptable exertional dyspnea because of an elevation in dead space ventilation. Operation in this circumstance is performed to reperfuse lung tissue, to reestablish a more normal ventilation, perfusion relationship (thereby reducing minute ventilatory requirements during rest and exercise), and to preserve the integrity of the contralateral circulation. If not previously implanted, an inferior vena caval filter is routinely placed several days in advance of the operation.

Guiding Principles of the Operation

There are several guiding principles for the operation. It must be bilateral because, for pulmonary hypertension to be a major factor, both pulmonary arteries must be substantially involved. The only reasonable approach to both pulmonary arteries is through a median sternotomy incision. Historically, there were many reports of unilateral operation, and occasionally this is still performed, in inexperienced centers, through a thoracotomy. However, the unilateral approach ignores the disease on the contralateral side, subjects the patient to hemodynamic jeopardy during the clamping of the pulmonary artery, and does not allow good visibility because of the continued presence of bronchial blood flow. In addition, collateral channels develop in chronic thrombotic hypertension not only through the bronchial arteries but also from diaphragmatic, intercostal, and pleural vessels. The dissection of the lung in the pleural space via a thoracotomy incision can therefore be extremely bloody. The median sternotomy incision, apart from providing bilateral access, avoids entry into the pleural cavities and allows the ready institution of cardiopulmonary bypass.

Cardiopulmonary bypass is essential to ensure cardiovascular stability when the operation is performed and to cool the patient to allow circulatory arrest. Very good visibility is required, in a bloodless field, to define an adequate endarterectomy plane and to then follow the pulmonary endarterectomy specimen deep into the subsegmental vessels. Because of the copious bronchial blood flow usually present in these cases, periods of circulatory arrest are necessary to ensure perfect visibility. Again, there have been sporadic reports of the performance of this operation without circulatory arrest. However, it should be emphasized that although endarterectomy is possible without circulatory arrest, a complete endarterectomy is not. We always initiate the procedure without circulatory arrest, and a variable amount of dissection is possible before the circulation is stopped, but never complete dissection. The circulatory arrest periods are limited to 20 minutes, with restoration of flow between each arrest. With experience, the endarterectomy usually can be performed with a single period of circulatory arrest on each side.

A true endarterectomy in the plane of the media must be accomplished. It is essential to appreciate that the removal of visible thrombus is largely incidental to this operation. Indeed, in most patients, no free thrombus is present; and on initial direct examination, the pulmonary vascular bed may appear normal. The early literature on this procedure indicates that thrombectomy was often performed without endarterectomy, and in these cases the pulmonary artery pressures did not improve, often resulting in death.

Surgical Technique

After a median sternotomy incision is made, the pericardium is incised longitudinally and attached to the wound edges. Typically the right heart is enlarged, with a tense right atrium and a variable degree of tricuspid regurgitation. There is usually severe right ventricular hypertrophy, and with critical degrees of obstruction, the patient's condition may become unstable with the manipulation of the heart. Anticoagulation is achieved with the use of beef-lung heparin sodium (400 units/kg, intravenously) administered to prolong the activated clotting time beyond 400 seconds. Full cardiopulmonary bypass is instituted with high ascending aortic cannulation and two caval cannulae. These cannulae must be inserted into the superior and inferior vena cavae sufficiently to enable subsequent opening of the right atrium. The heart is emptied on bypass, and a temporary pulmonary artery vent is placed in the midline of the main pulmonary artery 1 cm distal to the pulmonary valve. This will mark the beginning of the left pulmonary arteriotomy.

When cardiopulmonary bypass is initiated, surface cooling with both the head jacket and the cooling blanket is begun. The blood is cooled with the pump-oxygenator. During cooling a 10°C gradient between arterial blood and bladder or rectal temperature is maintained.[17] Cooling generally takes 45 minutes to

an hour. When ventricular fibrillation occurs, an additional vent is placed in the left atrium through the right superior pulmonary vein. This prevents atrial and ventricular distension from the large amount of bronchial arterial blood flow that is common with these patients. It is most convenient for the primary surgeon to stand initially on the patient's left side. During the cooling period, some preliminary dissection can be performed, with full mobilization of the right pulmonary artery from the ascending aorta. All dissection of the pulmonary arteries takes place intrapericardially, and neither pleural cavity should be entered. An incision is then 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 take-off of the middle lobe artery.

Any loose thrombus, if present, is now removed. 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. Therefore, to the inexperienced or cursory glance, the pulmonary vascular bed may well appear normal even in patients with severe chronic embolic pulmonary hypertension. 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.

When the patient's temperature reaches 20°C, the aorta is crossclamped and a single dose of cold cardioplegic solution (1L) is administered. Additional myocardial protection is obtained by the use of a cooling jacket. The entire procedure is now performed with a single aortic crossclamp period with no further administration of cardioplegic solution. A modified cerebellar retractor is placed between the aorta and superior vena cava. 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. It is rare that one 20-minute period for each side is exceeded. Although retrograde cerebral perfusion has been advocated for total circulatory arrest in other procedures, it is not helpful in this operation because it does not allow a completely bloodless field, and with the short arrest times that can be achieved with experience, it is not necessary.

Removing Thromboembolic Material

Any residual loose thrombotic debris encountered is removed. Then, a microtome knife is used to develop the endarterectomy plane posteriorly. Dissection in the correct plane is critical because if the plane is too deep the pulmonary artery may perforate, with fatal results, and if the dissection plane is not deep enough, inadequate amounts of the chronically thromboembolic material will be removed. Once the plane is correctly developed, a full-thickness layer is left in the region of the incision to ease subsequent repair. 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. This is why 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. Once the right-side 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.

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. 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 left-sided dissection is virtually analogous in all respects to that accomplished on the right. The duration of circulatory arrest intervals during the performance of the left-side dissection is subject to the same restriction as the right. After the completion of the endarterectomy, cardiopulmonary bypass is reinstituted and warming is commenced. Methylprednisolone (500 mg, intravenously) and mannitol (12.5 g, intravenously) are administered, and during warming a 10°C temperature gradient is maintained between the perfusate and body temperature. If the systemic vascular resistance level is high, nitroprusside is administered to promote vasodilatation and warming. The rewarming period generally takes approximately 90 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 right atrium is then opened and examined, unless prior to cardiopulmonary bypass, a negative "bubble" test was confirmed on transesophageal echocardiography. Otherwise, any intraatrial communication (present in about 20% of patients) is closed at this point. 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. 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. Myocardial cooling is discontinued once all cardiac procedures have been concluded. The left atrial vent is removed, and the vent site is repaired. All air is removed from the heart, and the aortic crossclamp is removed.

When the patient has rewarmed, cardiopulmonary bypass is discontinued. Dopamine hydrochloride is routinely administered at renal doses, and other inotropic agents and vasodilators are titrated as necessary to sustain acceptable hemodynamics. The cardiac output is generally high, with a low systemic vascular resistance. Temporary atrial and ventricular epicardial pacing wires are placed. Despite the duration of extracorporeal circulation, hemostasis is readily achieved, and the

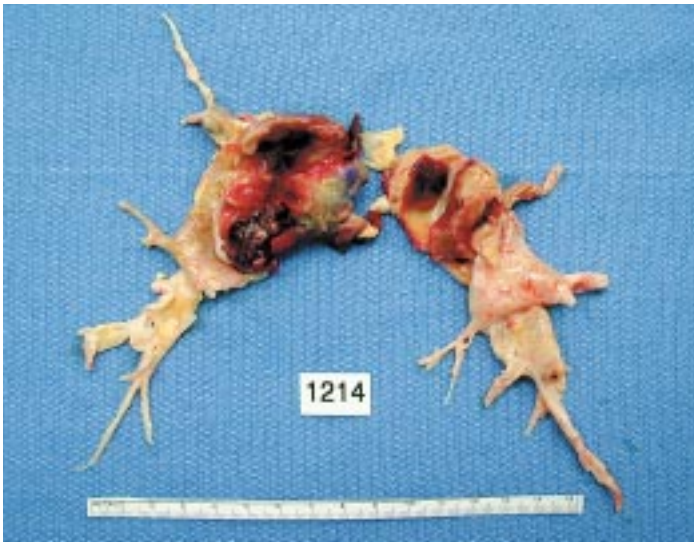

Fig. 1—Surgical specimen removed from right and left pulmonary arteries. Evidence of fresh thrombus indicates type I disease. Note that removal of only the fresh material leaves a large amount of disease behind. The ruler measures 15 cm.

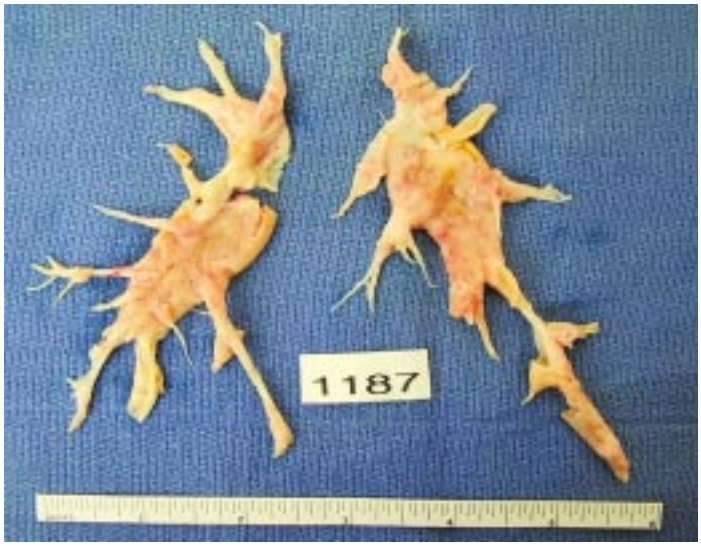

Fig. 2—Surgical specimen removed from right and left pulmonary arteries indicating type II disease. Note the extent of dissection down to the tail end of each branch. The ruler measures 6 inches.

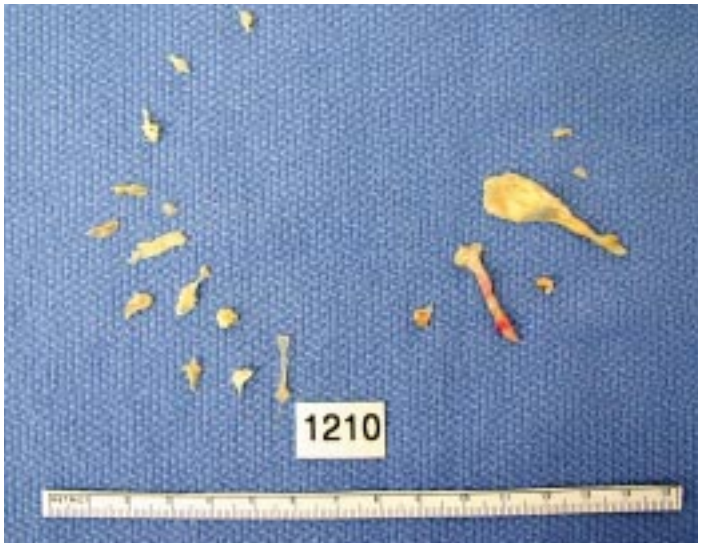

Fig. 3—Surgical specimen removed from right and left pulmonary arteries. In this patient the dissection plane was raised at each segmental level. The ruler measures 15 cm.

administration of platelets or coagulation factors is generally unnecessary. Wound closure is routine. A vigorous diuresis is usual for the next few hours, also a result of the previous systemic hypothermia.

Disease Classification: Four Types

There are four broad types of pulmonary occlusive disease related to thrombus that have been described by our group [15]:

1) Type I disease (approximately 30% of cases of thromboembolic pulmonary hypertension; **Fig. 1**) refers to the situation in which major vessel clot is present and readily visible on the opening of the pulmonary arteries. As mentioned earlier, all central thrombotic material has to be completely removed before the endarterectomy.
2) In type II disease (approximately 60% of cases; **Fig. 2**), no major vessel thrombus can be appreciated. In these cases only thickened intima can be seen, occasionally with webs, and the endarterectomy plane is raised in the main, lobar, or segmental vessels.
3) Type III disease (approximately 10% of cases; **Fig. 3**) 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 III disease is most often associated with presumed repetitive thrombi from indwelling catheters (such as pacemaker wires) or ventriculoatrial shunts, and sometimes represents "burnt out" disease, where most of the embolic material has been reabsorbed.
4) Type IV disease does not represent classic chronic 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. Small-vessel disease may be unrelated to thromboembolic events ("primary" pulmonary hypertension) or occur in relation to thromboembolic hypertension as a result of a high flow or high-pressure state in previously unaffected vessels similar to the generation of Eisenmenger's syndrome. We believe that there may also be sympathetic "cross-talk" from an affected contralateral side or stenotic areas in the same lung.

Postoperative Care

Meticulous postoperative management is essential to the success of this operation. All patients are mechanically ventilated for at least 24 hours, and all patients are subjected to a maintained diuresis with the goal of reaching the patient's preoperative weight within 24 hours. Although much of the postoperative care is common to more ordinary open-heart surgery patients, there are some important differences. The electrocardiogram, systemic and pulmonary arterial and central venous pressures, temperature, urine output, arterial oxygen saturation,

chest tube drainage, and fluid balance are monitored. A pulse oximeter is used to continuously monitor peripheral oxygen saturation. Management of cardiac arrhythmias and output and treatment of wound bleeding are identical to other open-heart operations. In addition, higher minute ventilation is often required early after the operation to compensate for the temporary metabolic acidosis that develops after the long period of circulatory arrest, hypothermia, and cardiopulmonary bypass. Tidal volumes higher than those normally recommended after cardiac surgery are therefore generally used to obtain optimal gas exchange. The maximum inspiratory pressure is maintained below 30 cm of water if possible. Although we used to believe that prolonged sedation and ventilation were beneficial and led to less pulmonary edema, subsequent experience has shown this not to be so. Extubation should be performed on the first postoperative day, if possible.

A specific complication that occurs in most patients to some degree is localized pulmonary edema, or the "reperfusion response." Reperfusion injury is defined as a radiologic opacity seen in the lungs within 72 hours of pulmonary endarterectomy.

Diuresis. Patients have considerable positive fluid balance after operation. After hypothermic circulatory arrest, patients initiate an early spontaneous aggressive diuresis for unknown reasons, but this may, in part, be related to the increased cardiac output related to a now lower PVR level. This should be augmented with diuretics, however, with the aim of returning the patient to the preoperative fluid balance within 24 hours of operation. Because of the increased cardiac output, some degree of systemic hypotension is readily tolerated. Fluid administration is minimized, and the patient's hematocrit level should be maintained above 30% to increase oxygen carrying capacity and mitigate the pulmonary reperfusion phenomenon.

Arrhythmias. The development of atrial arrhythmias, at approximately 10%, is no more common than that encountered in patients who undergo other types of nonvalvular heart surgery. The small, inferior atrial incision, away from the conduction system of the atrium or its blood supply, may be helpful in the reduction of the incidence of these arrhythmias.

Transfusion. Despite the requirement for the maintenance of an adequate hematocrit level, with careful blood conservation techniques used during operation, transfusion is required in a minority of patients.

Anticoagulation. A Greenfield filter is usually inserted before operation, to minimize recurrent pulmonary embolism after pulmonary endarterectomy. However, if this is not possible, it can also be inserted at the time of operation. If the device is to be placed at operation, radiopaque markers should be placed over the spine that correspond to the location of the renal veins to allow correct positioning. Postoperative venous thrombosis prophylaxis with intermittent pneumatic compression devices is used, and the use of subcutaneous heparin is begun on the evening of surgery. Anticoagulation with warfarin is begun as soon as the pacing wires and mediastinal drainage tubes are removed, with a target international normalized ratio of 2.5 to 3.

Complications

Patients are subject to all complications associated with open heart and major lung surgery (arrhythmias, atelectasis, wound infection, pneumonia, mediastinal bleeding, etc.) but also may develop complications specific to this operation. These include persistent pulmonary hypertension, reperfusion pulmonary response, and neurologic disorders related to deep hypothermia.

Persistent Pulmonary Hypertension. The decrease in PVR level usually results in an immediate and sustained restoration of pulmonary artery pressures to normal levels, with a marked increase in cardiac output. In a few patients, an immediately normal pulmonary vascular tone is not achieved, but an additional substantial reduction may occur over the next few days because of the subsequent relaxation of small vessels and the resolution of intraoperative factors such as pulmonary edema. In such patients, it is usual to see a large pulmonary artery pulse pressure, the low diastolic pressure indicating good runoff, and yet persistent pulmonary arterial inflexibility still resulting in a high systolic pressure.

There are a few patients in whom the pulmonary artery pressures do not resolve substantially. We do operate on some patients with severe pulmonary hypertension but equivocal embolic disease. Despite the considerable risk of attempted endarterectomy in these patients, since transplantation is the only other avenue of therapy, there may be a point when it is unlikely that a patient will survive until a donor is found. In our most recent 500 patients, more than one third of perioperative deaths were directly attributable to the problem of inadequate relief of pulmonary artery hypertension. This was a diagnostic rather than an operative technical problem. Attempts at pharmacologic manipulation of high residual PVR levels with sodium nitroprusside, epoprostenol sodium, or inhaled nitric oxide are generally not effective. Because the residual hypertensive defect is fixed, it is not appropriate to use mechanical circulatory support or extracorporeal membrane oxygenation in these patients if they deteriorate subsequently.

The Reperfusion Response. A specific complication that occurs in most patients to some degree is localized pulmonary edema, or the reperfusion response. Reperfusion injury is defined as a radiologic opacity seen in the lungs within 72 hours of pulmonary endarterectomy. This unfortunately loose definition may therefore encompass many causes, such as fluid overload and infection. True reperfusion injury that directly adversely impacts the clinical course of the patient now occurs in approximately 10% of patients. In its most dramatic form, it occurs soon after operation {within a few hours) and is associated with profound desaturation. Edema-like fluid, sometimes with a bloody tinge, is suctioned from the endotracheal tube. Frank blood from the endotracheal tube, however, signifies a mechanical violation of the blood-airway barrier that has occurred at operation and stems from a technical error. This complication should be managed, if possible, by identification of the affected area by bronchoscopy and balloon occlusion of the affected lobe until coagulation can be normalized.

One common cause of the reperfusion pulmonary edema is persistent high pulmonary artery pressures after operation when a thorough endarterectomy has been performed in certain areas, but there remains a large part of the pulmonary vascular bed affected by type IV change. However, the reperfusion phenomenon is often encountered in patients after a seemingly technically perfect operation with complete resolution of high

pulmonary artery pressures. In these cases the response may be one of reactive hyperemia, after the revascularization of segments of the pulmonary arterial bed that have long experienced no flow. Other contributing factors may include perioperative pulmonary ischemia and conditions associated with high permeability lung injury in the area of the now denuded endothelium. Fortunately, the incidence of this complication is very much less common now in our series, probably as a result of the more complete and expeditious removal of the endarterectomy specimen that has come with the large experience over the last few years.

Management of the Reperfusion Response. Early measures should be taken to minimize the development of pulmonary edema with diuresis, maintenance of the hematocrit levels, and the early use of peak end-expiratory pressure. Once the capillary leak has been established, treatment is supportive because reperfusion pulmonary edema will eventually resolve if satisfactory hemodynamics and oxygenation can be maintained. Careful management of ventilation and fluid balance is required. The hematocrit is kept high (32%-36%), and the patient undergoes aggressive diuresis, even if this requires ultrafiltration. The patient's ventilatory status may be dramatically position sensitive. The F_{IO_2} level is kept as low as is compatible with an oxygen saturation of 90%. A careful titration of positive end-expiratory pressure is carried out, with a progressive transition from volume-limited to pressure-limited inverse ratio ventilation and the acceptance of moderate hypercapnia. Infrequently, inhaled nitric oxide at 20 to 40 parts per million can improve the gas exchange. On occasion we have used extracorporeal perfusion support (extracorporeal membrane oxygenator or extracorporeal carbon dioxide removal) until ventilation can be resumed satisfactorily, usually after 7 to 10 days.

Delirium. Early in the pulmonary endarterectomy experience (before 1990), there was a substantial incidence of postoperative delirium. A study of 28 patients who underwent pulmonary endarterectomy showed that 77% experienced the development of this complication.[18] Delirium appeared to be related to an accumulated duration of circulatory arrest time of more than 55 minutes; the incidence fell to 11% with significantly shorter periods of arrest time.[19] With the more expeditious operation that has come with our increased experience, postoperative confusion is now encountered no more commonly than with ordinary open-heart surgery.

Results

More than 1575 pulmonary thromboembolism operations have been performed at UCSD Medical Center since 1970. Nearly 1400 have been completed since 1990, when the surgical procedure was modified as described earlier. The mean patient age in the last 1300 patients was 52 years, with a range of 8 to 85 years. There was a very slight male predominance. In nearly one third of these cases, at least one additional cardiac procedure was performed at the time of operation. Most commonly, the adjunct procedure was closure of a persistent foramen ovale or atrial septal defect (26%) or coronary artery bypass grafting (8%).

It is increasingly apparent that pulmonary hypertension caused by chronic pulmonary embolism is a condition that is underrecognized and carries a poor prognosis. Medical therapy is ineffective in prolonging life and only transiently improves the symptoms. The only therapeutic alternative to pulmonary thromboendarterectomy is lung transplantation.

Hemodynamic Results. A reduction in pulmonary pressures and resistance to normal levels and a corresponding improvement in pulmonary blood flow and cardiac output are generally immediate and sustained. In general, these changes can be assumed to be permanent. Before the operation, more than 95% of the patients are in NYHA functional class III or IV; at 1 year after the operation, 95% of patients remain in NYHA functional class I or II.[20, 21] In addition, echocardiographic studies have demonstrated that, with the elimination of chronic pressure overload, right ventricular geometry rapidly reverts toward normal. Right atrial and right ventricular enlargement regresses. Tricuspid valve function returns to normal within a few days as a result of restoration of tricuspid annular geometry after the remodeling of the right ventricle, and tricuspid repair is not therefore part of the operation.

Operative Morbidity. Severe reperfusion injury was the single most frequent complication in the UCSD series, occurring in 10% of patients. Some of these patients did not survive, and other patients required prolonged mechanical ventilatory support. A few patients were salvaged only by the use of extracorporeal support and blood carbon dioxide removal. Neurologic complications from circulatory arrest appear to have been eliminated, probably as a result of the shorter circulatory arrest periods now experienced, and perioperative confusion and stroke are now no more frequent than with conventional open-heart surgery. Early postoperative hemorrhage required reexploration in 2.5% of patients, and only 50% of patients required intra- or postoperative blood transfusion. Despite the prolonged operation, wound infections are relatively infrequent. Only 1.8% experienced the development of sternal wound complications, including sterile dehiscence or mediastinitis.

Deaths. In our experience, the overall mortality rate (30 days or in-hospital if the hospital course is prolonged) was 9% for the entire patient group, which encompasses a time span of 30 years. The mortality rate was 9.4% in 1989 and has been 5% to 7% for the more than 1300 patients who have undergone the operation since 1990. In the most recent series of 500 patients (June 1998 to July 2002), the mortality rate was 4.4%. We generally quote an operative risk of approximately 5%, but some patients predictably fall within a much higher risk. With our increasing experience and many referrals, we continue to accept the occasional patient who, in retrospect, was unsuitable for the procedure. We also accept patients in whom we know that the entire degree of pulmonary hypertension cannot be explained by the occlusive disease detected by angiography but feel that they will be benefited by operation, albeit at higher risk. Residual causes of death are operation on patients in whom thromboembolic disease was not the cause of the pulmonary hypertension (50%) and the rare case of reperfusion pulmonary edema that progresses to a respiratory distress syndrome of long standing, which is not reversible (25%).

Summary

It is increasingly apparent that pulmonary hypertension caused by chronic pulmonary embolism is a condition that is under-recognized and carries a poor prognosis. Medical therapy is inef-

fective in prolonging life and only transiently improves the symptoms. The only therapeutic alternative to pulmonary thromboendarterectomy is lung transplantation. The advantages of thromboendarterectomy include a lower operative mortality and excellent long-term results without the risks associated with chronic immunosuppression and chronic allograft rejection. The mortality for thromboendarterectomy at our institution is now in the range of 4.5%, with sustained benefit. These results are clearly superior to those for transplantation both in the short and long term.

Although pulmonary thromboendarterectomy is technically demanding for the surgeon, and requires careful dissection of the pulmonary artery planes and the use of circulatory arrest; excellent short- and long-term results can be achieved. Successive improvements in operative technique developed over the last four decades allow pulmonary endarterectomy to be offered to patients with an acceptable mortality rate and excellent anticipation of clinical improvement. With this growing experience, it has also become clear that unilateral operation is obsolete and that circulatory arrest is essential. The primary problem remains that this is an under-recognized condition. Increased awareness of both the prevalence of this condition and the possibility of a surgical cure should avail more patients of the opportunity for relief from this debilitating and ultimately fatal disease.

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