Pulmonary Thromboendarterectomy

78

Michael M. Madani and Jill R. Higgins

High Yield Facts

- Pulmonary thromboendarterectomy (PTE) is a technically difficult operation that involves performing a complete endarterectomy of the pulmonary arteries under profound hypothermia and circulatory arrest.
- The operation reduces right ventricular impedance, improves perfusion of ventilated lung, and is the definitive treatment for chronic thromboembolic pulmonary hypertension (CTEPH).
- Thromboembolic pulmonary hypertension is underrecognized even in the Western world; although the true incidence of this disease is unknown and hard to establish, it is estimated that roughly abut 4–5% of patients with acute PE will go on to develop CTEPH.
- More recently, advancement of surgical techniques, along with the development of new instruments, have allowed us to offer and perform a complete endarterectomy even in patients with isolated segmental and subsegmental disease.
- For those who are determined inoperable by an expert center, due to the location of the obstructive disease or significant comorbidities, newer percutaneous interventions such as Balloon Pulmonary Angioplasty (BPA) may offer a potential treatment strategy.
- Although BPA is in its infancy, and requires multiple sessions, there are encouraging early results from a few centers in Japan, Europe, and the U.S.
- Most recently, a minimally invasive approach has been developed to avoid sternotomy in highly selected patients. The procedure is currently only offered to patients with more proximal disease, suitable anatomy for mini-anterior thoracotomy, and to those with no need for concomitant procedures.

Introduction

Acute pulmonary thromboembolism and its chronic sequelae are significant causes of morbidity and mortality. However, the chronic disease process that can occur, is often underdiagnosed due to the non-specific nature of the two major symptoms, dyspnea on exertion and fatigue. Furthermore, physical findings may be elusive until symptoms of right heart failure have developed. The condition carries a high morbidity and long-term prognosis is quite poor, if the patient remains untreated [1]. Pulmonary thromboendarterectomy (PTE) or pulmonary endarterectomy (PEA) is the definitive treatment for chronic thromboembolic pulmonary hypertension (CTEPH) [2, 3]. Although, acute pulmonary embolism is one of the more common cardiovascular diseases, CTEPH is significantly underdiagnosed, and as a result PTE remains an uncommon procedure. The disease is characterized by intraluminal thrombus organization, fibrotic scar tissue-like stenosis, and vascular remodeling of the unaffected pulmonary vessels. PTE or PEA is an operation that is curative for patients with CTEPH [2, 3].

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PTE is a technically demanding operation that is currently performed in a few select centers around the world. Proper patient selection, meticulous surgical technique, and careful postoperative management have contributed to the excellent outcomes of this operation [2–4]. A true endarterectomy (not an embolectomy) of all affected segments of the lung is essential to open up all affected areas of the pulmonary vasculature. The procedure, when performed successfully at an experienced center, significantly improves pulmonary hypertension, right ventricular dysfunction, tricuspid regurgitation, and limits retrograde expansion of thromboembolic material. It also improves perfusion of the ventilated lung, minimizing dead-space ventilation, and prevents arteriopathic changes in the remaining patent small pulmonary vessels. Here, we describe the surgical procedure as it is performed at the University of California San Diego (UCSD) Cardiovascular Institute. To date, UCSD has performed over 4000 PTEs, which is the largest volume of PTE surgeries in the world. It is this experience that forms the foundation of this chapter.

Indications

When the diagnosis of thromboembolic pulmonary hypertension has been confirmed, 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 [5] pointed out that there were three major reasons for considering thromboendarterectomy: hemodynamic, alveolo-respiratory, and prophylactic. The hemodynamic 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, regardless of the severity of pulmonary hypertension. 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. 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 7 to 88 years. A typical patient will have a severely elevated pulmonary vascular resistance (PVR) level at rest, absence of significant comorbid disease unrelated to right heart failure, and the appearances of chronic thrombi on angiogram that appear to be relatively in balance with the measured PVR level. Exceptions to this general rule occur in symptomatic patients who exhibit pulmonary hypertension only with exercise.

Although most patients have a PVR level of less than 1000 dynes/s/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 (approximately 20% in our practice) have a level of PVR in excess of 1000 dynes/s/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, as long as there is evidence of thromboembolic disease that correlates with the degree of pulmonary hypertension.

There is a subgroup of patients who have a PVR level that is normal at rest, although elevated with minimal exercise. The condition is generally referred to as chronic thromboembolic disease or CTED. This is usually a young patient with significant thromboembolic disease and on occasion unilateral pulmonary artery occlusion with unacceptable exertional dyspnea because of an elevation in dead space ventilation. Operation in this circumstance is performed not only to re-perfuse lung tissue, but to reestablish a more normal ventilation perfusion relationship (thereby reducing minute ventilatory requirements during rest and exercise), and also to preserve the integrity of the contralateral circulation and prevent chronic arterial changes associated with long-term exposure to pulmonary hypertension. Inferior vena cava (IVC) filter insertion prior to surgery used to be routine in many centers. It has not been formally studied and has been abandoned by leading centers. IVC filter was not shown to increase long term survival [6].

Operation Principles

There are several guiding principles for the operation. Surgical treatment and endarterectomy must be bilateral because this is a bilateral disease in the vast majority of our patients. Typically, PTE is performed through a median sternotomy. The median sternotomy incision, apart from providing bilateral access, avoids entry into the pleural cavities, and allows the ready institution of cardiopulmonary bypass. More recently, a minimally invasive approach has been developed. This is a highly specialized operative technique that requires the combined skillset of an experienced minimally invasive surgeon, and an experienced PTE surgeon. This is not an approach for the novice surgeon. It involves cardiopulmonary bypass and circulatory arrest, with direct access to the pulmonary arteries via mini anterior thoracotomy incisions, without the use of a cross-clamp.

Cardiopulmonary bypass is essential to ensure cardiovascular stability when the operation is performed and to cool the patient to allow circulatory arrest. Excellent 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. Circulatory arrest is essential in these patients in order to create a bloodless field. These patients typically have copious bronchial blood flow, which can obscure the view of distal vessels in the absence of circulatory arrest. 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 min, 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. Circulatory arrest allows for a complete endarterectomy without increasing any potential risk of the procedure [7].

A true endarterectomy in the plane of the intima/media must be accomplished. It is essential to appreciate that the removal of visible thrombus is largely incidental to this operation. 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 with the resultant death of the patient.

Technical Aspects

The patient is laid supine on the operating table and appropriate monitoring lines, including a radial as well as a femoral arterial line, and a pulmonary artery catheter are placed. A transesophageal echocardiogram is also placed by the anesthesiologist. Brain activity is monitored by electroencephalogram, and a cooling jacket is placed around the head. The body's temperature is measured by use of bladder temperature, rectal temperature, as well as tympanic temperature. Then, once the patient is prepped and draped, a median sternotomy is made. It is important to ensure excellent hemostasis as these patients typically have enlarged and thin-walled venous collaterals and high CVP. In addition, having excellent hemostasis helps with the exposure and dissection of pulmonary vessels, which lie deep within the mediastinum. The pericardium is then incised longitudinally and attached to the wound edges. Full cardiopulmonary bypass is instituted with high ascending aortic cannulation and 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.

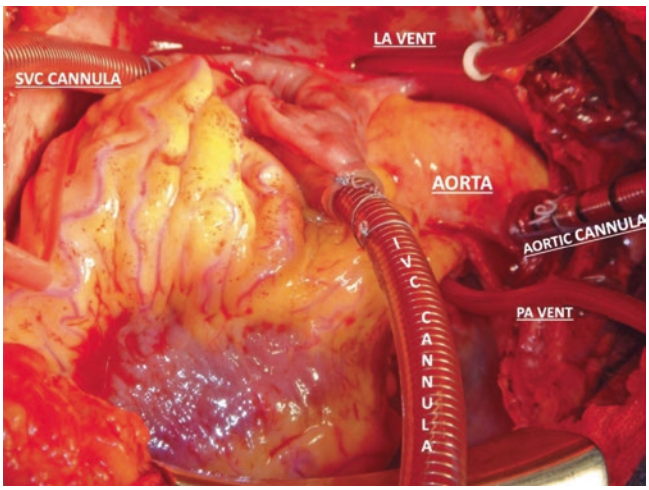

Fig. 78.1 Surgical field while the patient is cooling on cardiopulmonary bypass. The surgeon stands on the left side of the patient to start the endarterectomy of the right lung

The patient is then cooled to 20 °C with the pump oxygenator; surface cooling is performed with a cooling blanket as well as the head cooling jacket. Once ventricular fibrillation occurs, an additional vent is placed in the left atrium via the right superior pulmonary vein. Patients with CTEPH develop a large amount of bronchial collaterals, and as such it is important to ensure the left ventricle remains empty during the cooling process. 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 (Fig. 78.1).

The surgeon then waits until body's core temperature has reached about 20 °C, as measured by both bladder and rectal temperatures. The approach to the right pulmonary artery is from the left side and it is usually 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 with a tourniquet applied circumferentially. Care is taken to avoid any injury to the phrenic nerve. 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 DelNido 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, which surrounds 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. A modified cerebellar retractor, which has its tips filed, or a Madani PTE retractor is then used to expose the pulmonary artery between the aorta and the superior vena cava (Fig. 78.2).

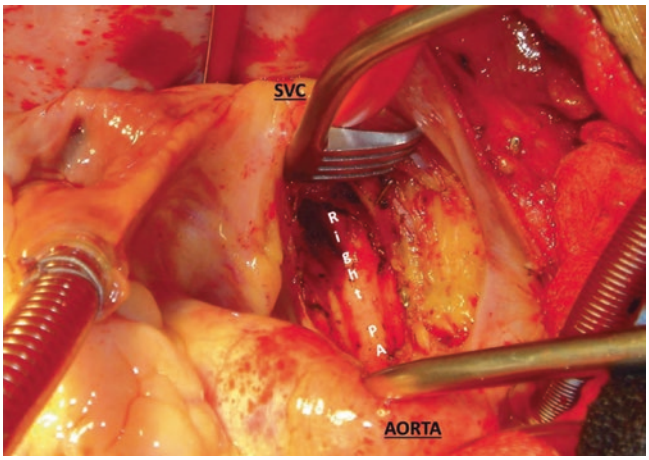

Fig. 78.2 Exposure of right pulmonary artery (PA)

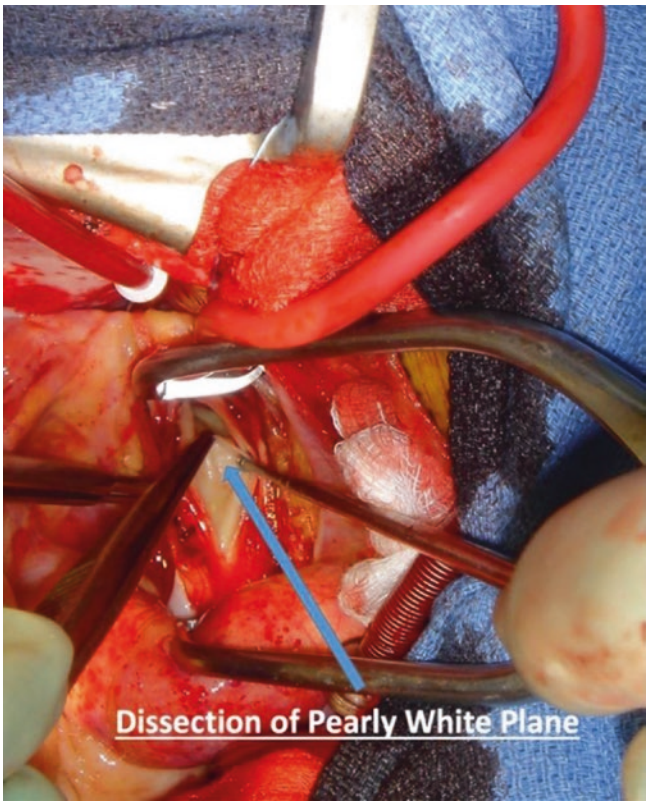

Fig. 78.3 Right pulmonary artery endarterectomy

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 important to recognize, however, an embolectomy without subsequent endarterectomy is quite ineffective. Furthermore, in most patients with chronic thromboembolic hypertension, direct examination of the pulmonary vascular bed at operation generally shows no obvious embolic material. Occasionally the bronchial circulation and back-bleeding is not excessive, the endarterectomy plane may then 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 (Fig. 78.3) 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. It is essential to identify the correct plane of dissection, which in most cases is between the intima and the media. A plane too deep will result in excessive thinning of the artery with the risk of tear in the distal branches, and a plane too shallow will result in incomplete endarterectomy.

The dissection is then carried out with the retractor in place. Circulatory arrest is then initiated, and the patient undergoes exsanguination into the reservoir. All monitoring lines to the patient are turned off to prevent aspiration of air. Snares are tightened around the cannulae in the superior and inferior venae cavae. It is rare to exceed a 20-min period for each side. The endarterectomy is then performed with an eversion technique. Because the vessel is everted and segmental and subsegmental branches are being worked on, a perforation here will become completely inaccessible and invisible later. This is why excellent visualization in an absolutely 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.

When only segmental and subsegmental disease is present, it is difficult to clearly visualize and perform a complete endarterectomy. In such distal cases, the plane of dissection is started more proximally in the normal plane of intima and/or occasionally media, and this plane of dissection is then carefully followed until the thickened fibrous tissue is reached. This technique can be quite challenging for the inexperienced surgeon as the remaining arterial tissue is quite thin and a tear through the arterial tissue is not inconsequential. A full endarterectomy is then completed in each of the segmental/subsegmental branches where disease is present. On occasion, circulatory arrest times of more than 20 min are required. In these situations, at about 20 min the circulation is restarted for about 10 min, at which time a second circulatory arrest is initiated, and the endarterectomy is completed. In most patients, the mixed venous oxygenation

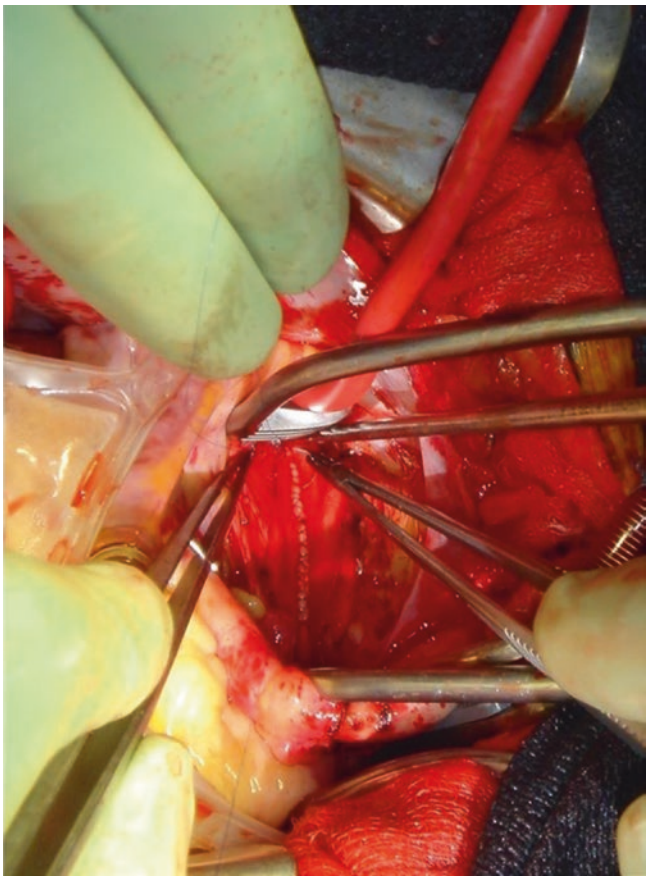

Fig. 78.4 Closure of right pulmonary artery

saturation is well above 90%, with 10 min of circulation. Once the right-sided endarterectomy is completed, circulation is restarted while maintaining a cold core body temperature. The pulmonary arteriotomy is then repaired with a continuous 6-0 polypropylene suture in a double-layer running fashion (Fig. 78.4).

After 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 similar to the right side and 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, which remains wrapped within a cooling jacket, is now retracted using a mesh-like net retractor shown in (Fig. 78.5).

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

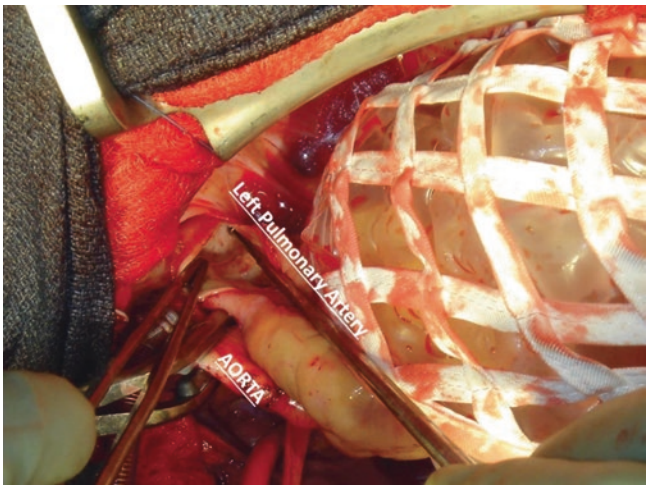

Fig. 78.5 Exposure of the left pulmonary artery

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 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.

After completion of the endarterectomy, cardiopulmonary bypass is reinstituted, and warming is commenced. The rewarming period generally takes approximately 90–120 min but varies according to the body mass of the patient. Once the left pulmonary arteriotomy has been sutured closed, the heart is returned to its normal position. If other cardiac procedures are required, such as closure of patent foramen ovale/atrial septal defect (ASD), coronary artery bypass grafting, mitral, or aortic valve surgery, these are conveniently performed during the systemic rewarming period. The most common concomitant procedure is closure of foramen ovale or ASDs, followed by coronary bypass surgery.

Although tricuspid valve regurgitation is invariable in these patients and is often moderate to severe, tricuspid valve repair is not necessary unless there is an anatomic abnormality with the valve leaflets, chords, or overall structure. Tricuspid regurgitation secondary to annular dilation is typically left alone as right ventricular remodeling occurs within a few days, with the return of tricuspid competence. However, in cases with severe annular dilation, with annular measurement of over 4–4.5 cm, it may be advisable to proceed with tricuspid annuloplasty to prevent potential recurrence in the future. 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. We typically use three 24 French soft mediastinal chest tubes, one of which is placed in the posterior pericardium as pericardial effusion posteriorly can occur. These drains are typically kept in place until drainage is below 200 cm^3/day and the patient is ambulatory. Wound closure is routine and similar to other cardiac procedures.

Figure 78.6 is surgical 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 specimen in Fig. 78.6 is characterized as level II disease on the right side and level III on the left side, according to the UCSD surgical classification.

There are four levels of pulmonary occlusive disease related to thrombus that can be appreciated, with level 0 (zero) symbolizing no evidence of chronic thromboembolic disease. Furthermore, the presence or absence of fresh thromboembolic material does not have any bearing on the classification. In other words, presence of stasis clot would no longer upgrade the surgical level of the disease. Table 78.1 summarizes the UCSD intra-operative classification.

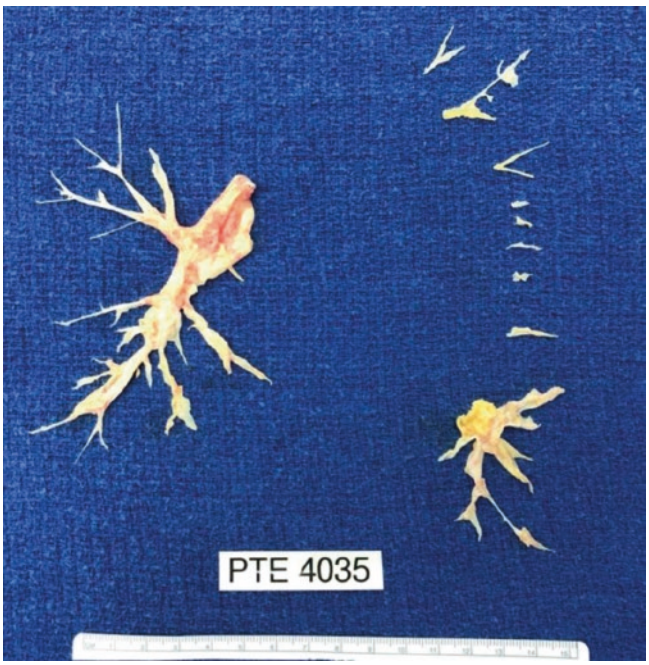

Fig. 78.6 Specimen removed from a patient indicating Level II disease on the right and Level III disease on the left

Table 78.1 UCSD CTEPH disease surgical classification

Level 0: No evidence of chronic thromboembolic disease in either lung
Level I: Chronic thromboembolic disease in the main pulmonary arteries
Level IC: Complete occlusion of one main pulmonary artery with chronic thromboembolic disease
Level II: Chronic thromboembolic disease starting at the level of lobar arteries, or in the main descending pulmonary arteries
Level III: Chronic thromboembolic disease starting at the level of the segmental arteries
Level IV: Chronic thromboembolic disease starting at the level of the subsegmental arteries

Postoperative care is similar to other cardiac procedures; and most patients are typically extubated within the first 48 h. The typical intensive care unit stay is about 2–3 days, and the average hospital stay is around 10–12 days. All patients undergo routine echocardiogram, V/Q scanning, as well as rest and exercise measurements of oxygen as a requirement prior to discharge. They will remain on lifelong anticoagulation using warfarin with a target international normalized ratio of about 3–4. Pulmonary endarterectomy can be curative for a vast majority of patients with excellent long-term outcomes. Our current 30-day and in-hospital and mortality rate is about 2% for all patients, and the current 10-year survival is about 75%. Most patients enjoy significantly improved hemodynamic numbers, and the majority will return to NYHA Class I/II functional level.

Surgical Advancements

With the advent of newly designed surgical instruments and retractor, we are now able to visualize distal pulmonary vasculature better and are able to remove disease that may be limited to only segmental and/or subsegmental vessels. By utilizing the techniques described above, we are now able to offer surgery to patients with distal disease, whom we may have turned down in the past. Figure 78.7 shows the surgical instruments used in PTE surgery at UC San Diego.

Also, more recently, the team at UCSD sought out to determine if a minimally invasive approach to PTE surgery was possible. Experiments were performed on multiple cadavers, which proved feasibility of performing a full endarterectomy into distal segmental and subsegmental arteries via mini anterior thoracotomy incisions, while providing adequate exposure. Using a preoperative computed tomography scan for surgical planning, the procedure is performed

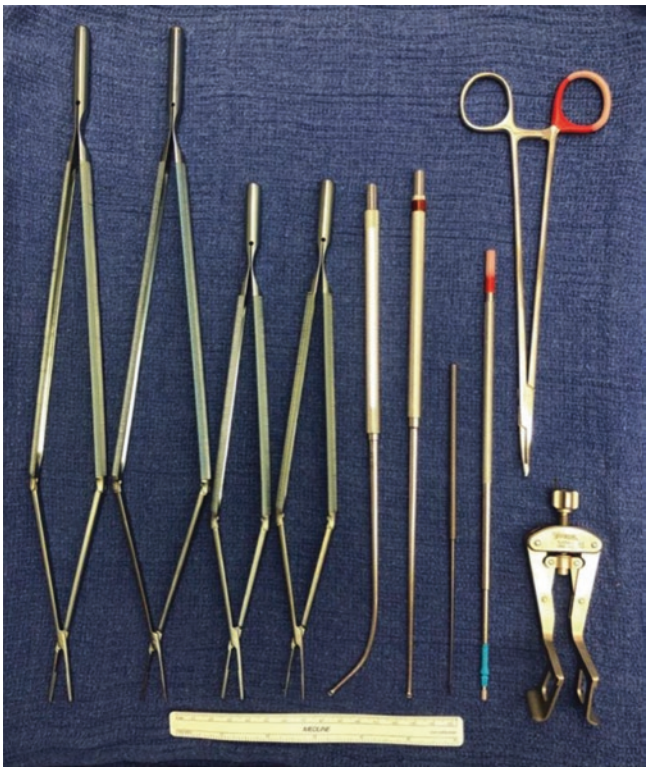

Fig. 78.7 Surgical instruments used during pulmonary thromboendarterectomy (PTE) surgery. From left to right, Madani double action PTE forceps, Jamieson suction/dissectors, microtome dissector, fine beaver blade, a "mini-peanut", and Madani PTE retractor

utilizing the second, or the third intercostal space through bilateral or unilateral mini anterior thoracotomies approximately 4–5 cm in length. The ideal location of the incisions is both high enough for central aortic cannulation, yet low enough for access to the pulmonary arteries. The arterial cannula is placed centrally in the ascending aorta, and venous cannulae in the femoral vein, right atrium and/or right internal jugular vein. For all patients, cardioplegia and cross-clamp were not used for purposes of simplification and to maximize space. An aortic root vent is intermittently utilized just prior to going back on cardiopulmonary bypass with each circulatory arrest. Pulmonary artery and left atrial vents are used. The usual protocol for circulatory arrest and exposure of the pulmonary arteries were used. Although in its infancy, with careful patient selection, and an expert surgeon, it is possible to perform a complete endarterectomy with visualization equivalent to that of a sternotomy. The minimally invasive approach to PTE surgery is not recommended for the novice PTE or minimally invasive cardiac surgeon.

The most important surgical advance has been redefining the limits of distal endarterectomy. In expert centers, PTE surgery can be successfully performed in patients with distal disease. This is attributed to advances in technology, instruments, and surgical experience. A new surgical classification has been developed to reflect the level of disease (i.e. lobar, segmental, subsegmental), as opposed to the type of disease. (UCSD classification Table 78.1). Depending on the level of experience of a center, operability may vary from center to center. A new definition of expert center has been proposed and evaluates the following; surgical mortality <5%, surgical volume >50 cases per year, and the ability to perform segmental endarterectomy, and the ability of the center to offer all treatment modalities (pulmonary thromboendarterectomy [PTE], balloon pulmonary angioplasty [BPA], medical therapy) [6].

Since the initial reports in 2012 from Japan, BPA has become an increasingly important tool in our armamentarium for patients who are not considered good surgical candidates. The results have varied from center to center and may not be generalizable, but they do show improved hemodynamic after multiple sessions of therapy. The long-term results and the effect of remaining disease within the pulmonary vasculature remains somewhat unknown. BPA should be reserved for expert centers, in patients who are not surgical candidates [6]. Figures 78.8 and 78.9 show the treatment algorithm proposed by the World Symposium on Pulmonary Hypertension and the treatment algorithm currently used at UC San Diego [8].

It is important to note that each treatment strategy affects different areas of the pulmonary vasculature. Clearly pulmonary hypertension directed medical therapy acts at the microvasculature level, and has very little effect on the obstructive disease as a result of thromboembolic occlusion. Both surgery and BPA address the obstructive disease, with

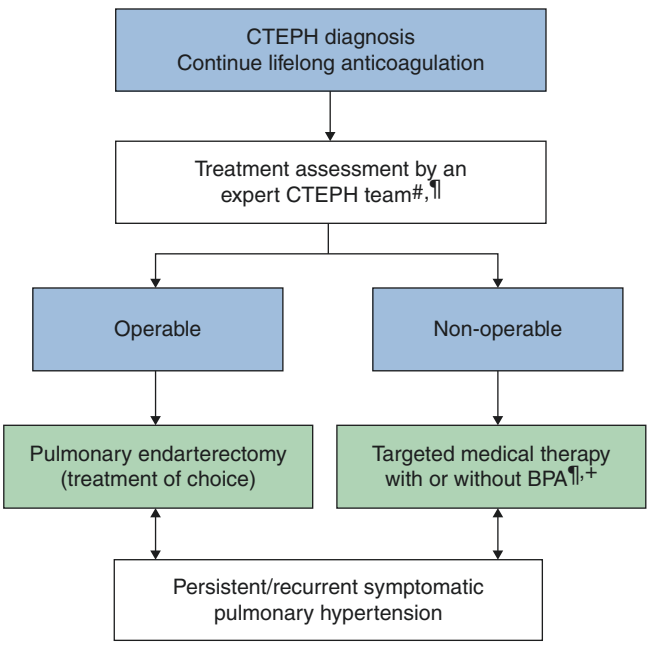

Fig. 78.8 Treatment algorithm proposed by the World Symposium on Pulmonary Hypertension [6]

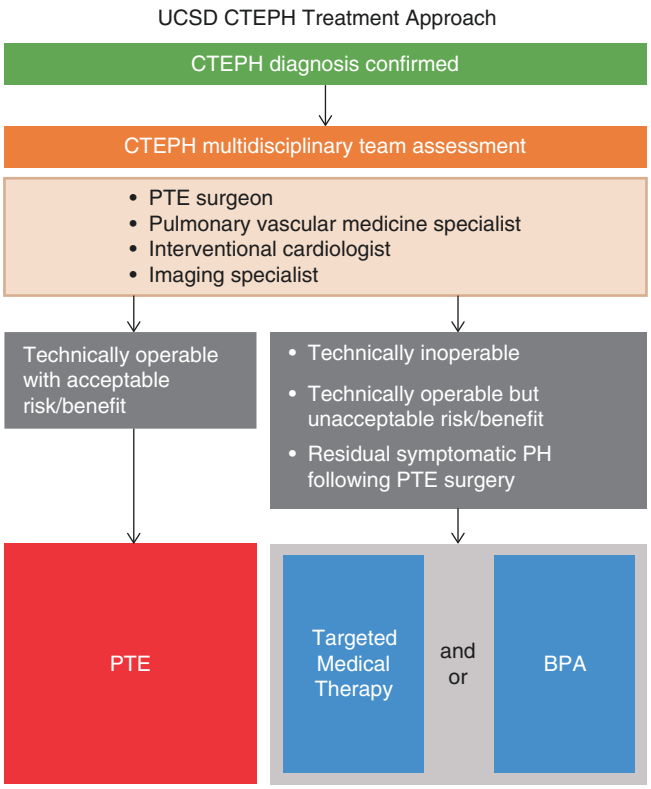

Fig. 78.9 Treatment algorithm used at UC San Diego for treatment of chronic thromboembolic pulmonary hypertension (CTEPH) [17]. *BPA* balloon pulmonary angioplasty, *PH* pulmonary hypertension, *PTE* pulmonary thromboendarterectomy

BPA generally addressing more distal disease at the subsegmental level and beyond. In experienced surgical hands disease starting at the segmental and even subsegmental level (Level III and IV), can be addressed and cleared. However, one can see how there may be a different treatment strategy at these distal levels depending on the center and the operators experience. Given the fact that the long-term results of BPA are unknown, and generally is a treatment that requires multiple sessions (generally 4–6 sessions), it should be reserved only for those patients who are not surgical candidates. Figure 78.10 is a cartoon indicating the different sites of action for each treatment strategy [8].

Results

More than 4000 pulmonary thromboendarterectomies have been performed at UCSD Medical Center since 1970. Most of these cases (over 3700) have been completed since 1990, when the surgical procedure was modified as described earlier in this chapter. The mean patient age in our group is about 51 years, with a range of 7–89 years. There is no gender difference. In nearly one-third of the 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%) [2, 4].

A reduction in pulmonary pressures and resistance to normal levels and a corresponding improvement in pulmonary

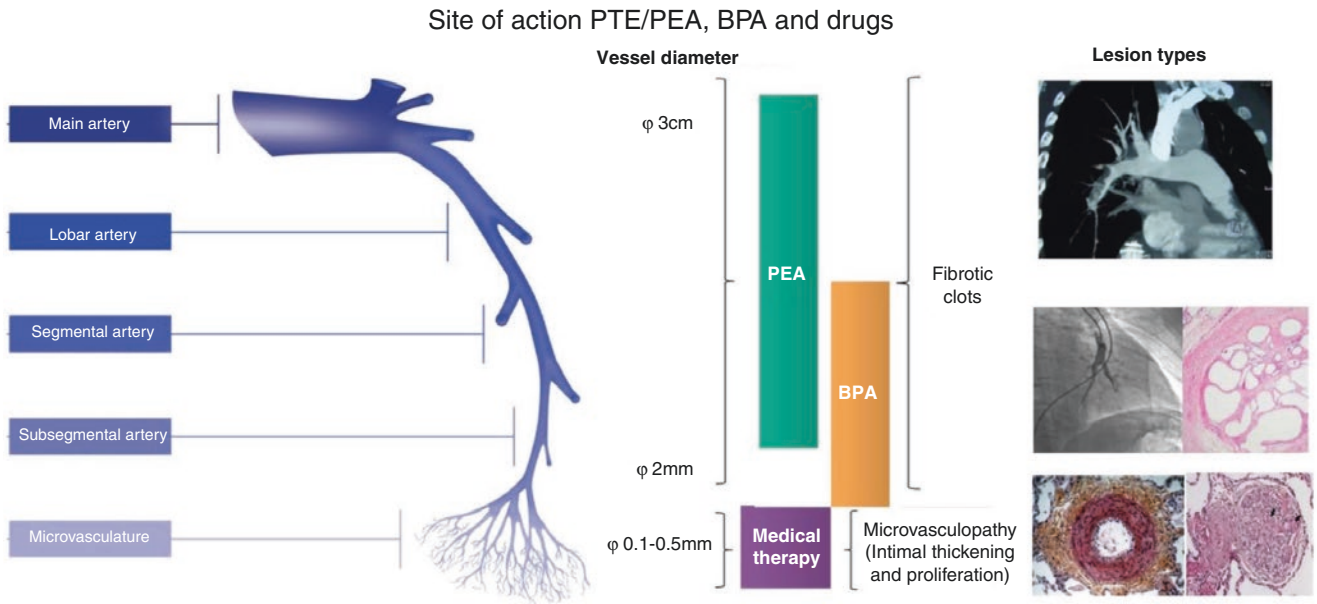

Fig. 78.10 Site of action for pulmonary thromboendarterectomy/pulmonary endarterectomy (PTE/PEA) surgery, balloon pulmonary angioplasty (BPA), and pulmonary hypertension (PH) directed medical therapy. Please note possible crossover of PTE/PEA and BPA at around segmental/subsegmental level. An experienced surgeon can clear disease that starts at distal segmental/proximal subsegmental level (Level IV) [8]

blood flow and cardiac output are generally immediate and sustained [2, 4]. In general, these changes can be assumed to be permanent. Whereas 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 [4]. 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. Neurological 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 [4, 7]. Early postoperative hemorrhage required reexploration in 2.5% of patients, and only 40% of patients required intra- or postoperative blood transfusion. Despite the prolonged operation, wound infections are relatively infrequent.

Mortality Rate

Mortality rate is inversely related to the surgeon's and center's experience. Cannon et al. [9] were able to show a significant difference in mortality rate after the first 500 operations. The difference was maintained over long-term follow-up. In our own experience at UC San Diego, the mortality rate was 9.4% in 1989 and then it remained at about half ranging 4–5% for the more than 2000 patients who had undergone the operation since 1990 until 2010. Looking at our most recent experience over the last 10 years, the mortality rate has been about 1–2%. 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 benefit from the operation, albeit at higher risk. With the recent availability of new medical therapies, as well as BPA, those patients with significant residual pulmonary hypertension can benefit from such therapy [10].

Other Post-operative Complications

Although rare, patients undergoing pulmonary endarterectomy are subject to all complications associated with open heart and major lung surgery (arrhythmias, atelectasis, wound infection, pneumonia, mediastinal bleeding) but also may develop complications specific to this operation. These include persistent pulmonary hypertension, reperfusion pulmonary edema, and airway bleeding or massive hemoptysis.

A specific complication that occurs in most patients to some degree is localized pulmonary edema, or the "reperfusion response." Reperfusion pulmonary edema is defined as a radiologic opacity seen in the lungs within 72 h of pulmonary endarterectomy. This unfortunately loose definition may therefore encompass many causes, such as fluid overload and infection. The complication typically occurs within 24–72 h after surgery, but rarely it can manifest as late as 1 week following surgery [8, 11–17].

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 pinkish bloody tinge, is suctioned from the endotracheal tube [8, 12]. 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, or occasionally may be related to bronchial collateral flow. 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. In certain circumstances, when one lung ventilation is not possible, extra-corporeal support may be required.

Conclusion

It is increasingly apparent that pulmonary hypertension caused by chronic pulmonary embolism is a condition that is under-recognized and carries a poor prognosis without any treatment. Medical therapy is generally ineffective in prolonging life and only available for patients who are not surgical candidates. Currently, the only therapeutic alternative to pulmonary thromboendarterectomy in those who have been deemed inoperable by an expert center, is BPA and rarely 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 2% with sustained benefit. These results are clearly superior to those for medical therapy or transplantation in both the short and the long term.

Although PTE 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, as long it is performed at expert centers [9]. The successive improvements in operative technique developed over the last four decades now allow pulmonary endarterectomy to be offered to patients with an acceptable mortality rate and excellent anticipation of clinical improvement.

The primary problem remains that this is an under-recognized and under-treated condition. The likely incidence

of CTEPH in the United States is at least 10 times the annual number of surgeries performed (approximately 400 per year). 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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