Chronic Thromboembolic Pulmonary Hypertension

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Abstract

Keywords

- pulmonary hypertension
- pulmonary embolism
- pulmonary endarterectomy

Chronic thromboembolic pulmonary hypertension occurs when acute thromboemboli fail to dissolve completely. The resulting fibrotic scar tissue within the pulmonary arteries is obstructive and eventually leads to right heart failure. Medical therapy for this condition is supportive, but surgery with pulmonary artery endarterectomy is curative, and carries a low mortality at experienced centers.

Objectives: Upon completion of this article, the reader will be able to (1) compare the epidemiology and pathophysiology of chronic thromboembolic pulmonary hypertension (CTEPH); (2) describe the latest diagnostic modalities and approach to evaluating patients with suspected chronic thromboembolic disease; and (3) illustrate the advancements in the surgical procedure, pulmonary thromboendarterectomy, currently available for patients with chronic thromboembolic disease and CTEPH.

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The estimates of the incidence of chronic thromboembolic disease after acute pulmonary embolism vary, with figures from 1 to 9%.^{1,2} However, it is likely that the development of this condition is more prevalent than realized; since most patients after acute thromboembolic episodes are not fol-

lowed serially with echocardiography (to assess the development of pulmonary hypertension), the diagnosis is often missed, and many patients (up to 50%) do not give a history of acute pulmonary embolism or deep venous thrombosis.³

If a figure of 5% of patients with acute embolic hypertension progressing to some degree of fibrotic obstructive disease is assumed, and then counting only patients who presented with symptomatic acute pulmonary emboli, ~25,000 patients would progress to chronic thromboembolic pulmonary hypertension in the United States each year.³ However, because many (if not most) patients diagnosed with chronic thromboembolic disease have no antecedent history of acute embolism, the true incidence of this disorder is probably much higher.

Regardless of the exact incidence or the circumstances, acute embolism and its chronic relation, fixed chronic thromboembolic occlusive disease, are both much more common than generally appreciated and are significantly underdiagnosed.

Although pulmonary embolism is one of the most common cardiovascular diseases affecting Americans, pulmonary thromboendarterectomy for the treatment of this form of pulmonary hypertension remains an uncommon procedure; this is because the condition remains underdiagnosed, and the current success of pulmonary endarterectomy is not widely appreciated.

Twenty-five years ago, the operation was only performed at the University of California, San Diego (UCSD). However, with the increased awareness of the efficacy of this operation,

pulmonary endarterectomy is now being performed at many centers both within the United States and internationally, with good results.

Etiology

When the acute thrombus does not resolve, the remaining clot becomes fibrotic and incorporated into the pulmonary artery wall. The progressive occlusion of the pulmonary vasculature results in pulmonary hypertension and eventual right heart failure.

The reasons for the failure of emboli to dissolve are not fully understood and may be from a combination of factors including repetitive thrombi and embolization of already partially fibrotic material, or the volume of acute embolic material may simply overwhelm the lytic mechanisms. In addition, total occlusion of a major arterial branch may prevent lytic elements from reaching, and thus dissolving, the embolus completely, or the lytic mechanisms themselves may be abnormal. Some patients may have a hypercoagulable state with a propensity for thrombus formation.

After the clot becomes wedged in the pulmonary artery, and fails to lyse, then the clot may canalize, producing multiple small endothelialized channels separated by fibrous septa (i.e., bands and webs). Alternatively, complete fibrous organization of the clot may result, leading to a solid mass of dense fibrous connective tissue totally obstructing the arterial lumen.⁴

There are some other special circumstances. Chronic indwelling central venous catheters and pacemaker leads are sometimes associated with pulmonary emboli, probably due to repetitive micro thromboemboli. Rarely, tumor emboli occur from stomach, breast, and kidney malignancies, and right atrial myxomas may also fragment and embolize.³

A hypercoagulable state may be present in some patients, which may encourage embolization, lead to spontaneous thrombosis within the pulmonary vascular bed, or be responsible for proximal propagation of thrombus after an embolus.

The genesis of the resultant pulmonary vascular hypertension due to partial occlusion of vessels may be complex. The increased pressure and flow because of redirected pulmonary blood flow in previously normal pulmonary small vessels can create a vasculopathy similar to that seen in Eisenmenger's syndrome. Factors other than the simple hemodynamic consequences of redirected blood flow may also be involved in this process. For example, after a pneumonectomy, 100% of the right ventricular output flows to one lung, yet little increase in pulmonary pressure occurs, even after a decade's follow-up. In patients with thromboembolic disease, pulmonary hypertension can occur when less than 50% of the vascular bed is occluded. Thus, sympathetic neural connections, hormonal changes or both, might initiate pulmonary hypertension in the initially unaffected pulmonary vascular bed. This process can occur with the initial occlusion either being in the same or the contralateral lung. Since these changes are not operable or reversible, with the low morbidity and mortality currently achieved with pulmonary endarterectomy, we advise earlier operation, before these changes can occur.

Presentation

Because of the large area of the pulmonary vascular bed, usually more than 60% of the vasculature must be occluded before pulmonary hypertension occurs in the absence of exercise.³ There are no signs or symptoms specific to chronic thromboembolism, and the diagnosis may be missed if the patient is asymptomatic at rest. On exercising, with the attendant increase in cardiac output, even with lesser degrees of occlusion, the pulmonary artery pressures will increase.

The most common symptom then, as with all other causes of pulmonary hypertension, is exertional dyspnea, out of proportion to any abnormalities found on clinical examination. Syncope, or light-headedness during exertion, is another common symptom of pulmonary hypertension, generally occurring in patients with more advanced disease and higher pulmonary arterial pressures.³

Nonspecific chest pain occurs in about half of patients with more severe pulmonary hypertension. Hemoptysis can happen in all forms of pulmonary hypertension and probably results from abnormally dilated vessels distended by increased intravascular pressure. Peripheral edema, early satiety, and epigastric or right upper quadrant fullness or discomfort may develop as the right heart fails (cor pulmonale).

The physical signs of pulmonary hypertension are those of right heart failure. Initially, the jugular venous pulse is characterized by a large A-wave, and then, as the right heart fails, the V-wave becomes predominant. The right ventricle is usually palpable near the lower left sternal border, and pulmonary valve closure may be heard in the second intercostal space. The second heart sound is often narrowly split and varies normally with respiration; P2 is accentuated. A sharp systolic ejection click may be heard over the pulmonary artery. As the right heart fails, a right atrial gallop usually is present, and tricuspid insufficiency develops. Because of the large pressure gradient across the tricuspid valve in pulmonary hypertension, the murmur is high pitched and may not exhibit respiratory variation. These findings are different from those usually observed in tricuspid valvular disease. A murmur of pulmonic regurgitation may also be detected. A specific finding in many patients with chronic thromboembolic disease is a flow murmur at the back, probably from distended bronchial arteries.³

Investigations

Chest X-ray, electrocardiogram, and pulmonary function tests are not of great value in differentiating thromboembolic pulmonary hypertension from other forms of pulmonary hypertension, though these studies may indicate that pulmonary hypertension exists.

The chest X-ray in thromboembolic disease is often normal, but may show apparent vessel cutoffs of the lobar or segmental pulmonary arteries, or regions of oligemia, suggesting vascular occlusion. Enlargement of the right ventricle and the central pulmonary arteries may be seen. The left atrium and left ventricle will be of normal size.

The electrocardiogram demonstrates the findings of right ventricular hypertrophy, with right axis deviation and a dominant R-wave in V1.

Pulmonary function tests should be performed to exclude obstructive or restrictive intrinsic pulmonary parenchymal disease as the cause of the hypertension. In chronic pulmonary thromboembolic disease, these tests reveal minimal changes in lung volume and ventilation. Diffusing capacity is often reduced and may be the only abnormality on pulmonary function testing. Most patients are hypoxic with room air, with arterial oxygen tensions ranging between 50 and 83 torr. CO₂ tension is slightly reduced and is compensated by reduced bicarbonate. Dead space ventilation is increased.³

The most useful screening studies are two-dimensional surface echocardiography with Doppler imaging and radionuclide ventilation-perfusion scanning.

The echocardiogram typically demonstrates right atrial, right ventricular, and pulmonary artery enlargement, with right ventricular hypertrophy. The interventricular septum may be flattened and often will exhibit paradoxical motion, with encroachment of the right ventricular septum into the left ventricle. Varying degrees of tricuspid regurgitation are usually seen. Continuous wave Doppler of the tricuspid regurgitant jet will estimate the pulmonary artery systolic pressure (PAP = [tricuspid envelope]² × 4 + CVP). Because exercise typically increases the degree of pulmonary hypertension, echocardiography should be repeated with exercise whenever the disease is suspected, but the resting echocardiogram demonstrates only subtle abnormalities.

A ventilation-perfusion lung scan is the essential test for establishing the diagnosis of unresolved pulmonary thromboembolism. An entirely normal lung scan excludes the diagnosis of both acute and chronic thromboembolism. The usual lung scan pattern in most patients with primary pulmonary hypertension either is relatively normal or shows a diffuse nonuniform perfusion (moth-eaten appearance).³ When subsegmental or larger perfusion defects are noted on the scan, even if matched with ventilatory defects, pulmonary angiography is appropriate to confirm or rule out thromboembolic disease.

Computed tomography (CT) scanning is increasingly being used in the diagnosis. This can confirm occlusion of at least the main and lobar pulmonary arteries, but may miss smaller degrees of occlusion. A mosaic pattern of lung attenuation at CT is a sign of variable regional perfusion and will suggest chronic pulmonary thromboembolism.³

The signs seen in the pulmonary artery at CT are similar to those seen with conventional angiography.

At angiography, complete vessel cutoffs will show a "pouch defect," a concave appearance of the interrupted contrast.³ Organized thromboembolic lesions do not have the appearance of the intravascular filling defects seen with acute pulmonary emboli. Organized thrombi appear as unusual filling defects, webs, or bands, or completely thrombosed vessels that may resemble congenital absence of the vessel. Organized material along a vascular wall of a recanalized vessel produces a scalloped or serrated luminal edge.

Occasionally, calcification can be seen within chronic, non-fibrotic lesions. Because of both vessel-wall thickening and dilatation of proximal vessels, the contrast-filled lumen may appear relatively normal in diameter. Distal vessels demonstrate the rapid tapering and pruning characteristic of pulmonary hypertension.

At our institution, pulmonary angiography is always performed. Right heart catheterization done at the same time measures pulmonary artery pressures and calculates the pulmonary vascular resistance (PVR). This is helpful in planning the conduct of the operation and the likelihood of complete resolution. Furthermore, CT scans may miss lesions that are only present at subsegmental level.

Both MRI and CT are additive to the angiogram and need not be performed if the angiogram is negative, or clearly diagnostic. In general, the lower lobes of the lung are more involved with occlusion than the upper lobes, and the right lung more affected than the left, probably because of the larger blood flow to the right side and the lower lobes.

Although the term "surgically accessible" thromboembolic disease is often used, pulmonary hypertension because of emboli is almost always operable, in that the patient can be improved. Complete resolution, however, may depend on the pattern of thrombotic occlusion and the presence of a secondary vasculopathy. An important indicator is the degree of concomitance of the PVR and the amount of thromboembolic occlusion seen on angiogram. If the PVR seems out of balance to the amount of occlusion, this should not contraindicate operation, but may predict that complete restoration of flow to the pulmonary vascular bed may not be possible. The likely discordance is from the secondary small vessel disease described earlier.

In addition to pulmonary angiography, patients older than 45 years undergo coronary arteriography, and other cardiac investigation as necessary. If significant disease is found, additional cardiac surgery is performed at the time of pulmonary endarterectomy. Most patients who undergo operation are within New York Heart Association (NYHA) Class III or Class IV (III: marked limitation in activity due to symptoms, comfortable only at rest; IV: severe limitations, mostly bedbound patients).

A typical patient will have a severely elevated PVR at rest, the absence of significant comorbid disease unrelated to right heart failure, and the appearances of chronic thrombi on angiogram within reasonable balance with the measured PVR.

With the growth of our surgical experience, however, surgery is offered to patients with more distal thromboembolic disease that is contributing to, but probably not entirely responsible for, the patient's symptoms (Type III disease—see below) and, at the other end of the spectrum, those with advanced right-side cardiac failure with ascites and hepatic and renal dysfunction that is presumed reversible. There is no upper limit of PVR, pulmonary artery pressure, or right ventricular hypertrophy or failure that excludes a patient from operation at our institution.

If not previously implanted, an inferior vena cava filter is routinely placed prior to the operation, usually at the time of cardiac catheterization.

Operation

Pulmonary embolization rarely results in tissue necrosis because the bronchial circulation maintains viability of the lung parenchyma. Surgical endarterectomy thus allows the lung tissue to regain its function of gas exchange.

Early in our experience it was pointed out that there are three major reasons for considering thromboendarterectomy: hemodynamic, alveolar respiratory, and prophylactic. The hemodynamic goal is to ameliorate or prevent the right ventricular compromise caused by pulmonary hypertension. The respiratory objective is to improve lung 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, and the prevention of secondary small vessel changes in the remaining patent vessels.

The ages of the patients in our series ranged from 5 to 86 years. Although most patients have a PVR level in the range of 800 dynes/sec/cm⁵ and pulmonary artery pressures less than systemic, the hypertrophy of the right ventricle that occurs over time makes pulmonary hypertension to suprasystemic levels possible. Thus, many patients will have a PVR more than 1,000 dynes/sec/cm⁵, together with suprasystemic pulmonary artery pressures.

A rare patient might have a PVR that is normal at rest, although elevated with minimal exercise. This is usually a young patient with total unilateral pulmonary artery occlusion and exertional dyspnea because of an elevation in dead space ventilation. Operation in this situation is performed to reperfuse lung tissue, reduce 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 during cardiac catheterization.

There are several guiding principles for the operation.³ It must be bilateral because, for pulmonary hypertension to be a major factor, both pulmonary arteries must be substantially involved. The only practical approach to both pulmonary arteries is through a median sternotomy incision. The median sternotomy incision, apart from providing bilateral access, avoids entry into the pleural cavities (where there may be lung adhesions) and allows the 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. Excellent visibility is required, in a bloodless field, to define an adequate endarterectomy plane and then to 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. 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 important 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.

All dissection of the pulmonary arteries takes place intrapericardially, and neither pleural cavity is entered. 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 take-off of the middle lobe artery. Any loose thrombus, if present, is now removed, though in most patients, direct examination of the pulmonary vascular bed generally shows no obvious embolic material.

Dissection in the correct plane of the pulmonary artery 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. Circulatory arrest with drainage of the blood from the lungs provides the necessary visibility.

When the proper plane is entered, in the media of the vessel, the layer will strip easily, and the material left with the outer layers of the pulmonary artery will appear somewhat yellow. The ideal layer is marked with a pearly white plaque, which strips easily. There should be no residual yellow plaque. If the dissection is too deep, a reddish or pinkish color indicates the adventitia has been reached, and more superficial plane should be sought immediately.

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.

Once the right-side endarterectomy is completed, circulation is restarted with the patient still being cooled, and the arteriotomy is closed.

The left-side dissection also takes place within the pericardium, though this is mobilized to give access to the left pulmonary artery as far as the take-off of the left upper lobe. Again, the plane is found and the distal vessels freed of all obstructing material, using circulatory arrest.

After the 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 ~90 minutes but varies according to the body mass of the patient.

There are four broad types of pulmonary occlusive disease related to thrombus that can be appreciated, and we use the following classification:^{2,3} Type I disease (~10–15% of cases of thromboembolic pulmonary hypertension) is seen when major vessel clot is present and readily visible on the opening of the pulmonary arteries (→ **Fig. 1**). In Type II disease (~60% of cases), no major vessel thrombus is seen, only thickened intima, occasionally with webs, and the endarterectomy plane is raised in the main, lobar, or segmental vessels (→ **Figs. 2 and 3**). Type III disease (~15–25% of cases in our experience) presents the most challenging surgical situation

Fig. 1 Type 1 disease. Thrombus is present in the main pulmonary arteries.

Fig. 3 Type 2 disease. Typical type 2 disease. The thrombus seen in the right lower lobe is due to stasis.

(→**Fig. 4**). The disease here 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 often associated with presumed repetitive thrombi from indwelling catheters (such as pacemaker wires) or ventriculo-atrial shunts. Type IV disease does not represent thromboembolic pulmonary hypertension and is inoperable. In this entity, there is intrinsic small vessel disease, although secondary thrombus may occur because of stasis. Small-vessel disease may be unrelated to thromboembolic events (“primary” pulmonary hypertension) or occur in relation to thromboembolic hypertension because of a high-flow or high-pressure state in previously unaffected vessels similar to the generation of Eisenmenger’s syndrome. There may also be sympathetic

“cross-talk” from an affected contralateral side or stenotic areas in the same lung, as described earlier.

Although tricuspid valve regurgitation is invariable in these patients and is often severe, tricuspid valve repair is not performed unless there are separate structural abnormalities. 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. When the patient has rewarmed, cardiopulmonary bypass is discontinued.

Despite the duration of extracorporeal circulation, hemostasis is usually readily achieved, and the administration of blood, platelets, or coagulation factors is generally unnecessary. Wound closure is routine.

Fig. 2 Type 2. Here the right side is completely occluded with fibrotic scar.

Fig. 4 Type 3 disease. The plane must be raised at each segmental level.

Postoperative Care

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. Patients have considerable positive fluid balance after this prolonged operation with circulatory arrest. Fluid administration is minimized, and the patient's hematocrit level should be maintained above 30% to increase oxygen-carrying capacity and to mitigate against the pulmonary reperfusion phenomenon.

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 times the control value.

Complications

Complications of the procedure, apart from the usual complications sometimes seen after heart surgery, include residual pulmonary hypertension and reperfusion lung injury.

The decrease in PVR usually results in an immediate and sustained restoration of pulmonary artery pressures to normal levels, with a marked increase in cardiac output. However, there are some patients in whom some degree of residual pulmonary hypertension is permanent. Generally, this is seen in patients in whom the preoperative PVR and the degree of angiographic occlusion are discordant, with the presence of damage to previously unaffected small vessels, as described earlier.

Although an increased PVR preoperatively poses an increased risk, this statement should come with the caveat that it is postoperative PVR that signifies risk rather than preoperative PVR.⁵ Thus, every effort should be made to remove all obstructing material, even if this increases circulatory arrest time.

A specific complication that occurs in many patients to some degree is localized pulmonary edema, or the "reperfusion response." Reperfusion injury is defined as a radiological opacity seen in the lungs within 72 hours of pulmonary endarterectomy. This loose definition may thus encompass many causes, such as fluid overload and infection. Early measures should be taken to minimize the development of pulmonary edema with diuresis, maintenance of the hematocrit, and the early use of positive end-expiratory pressure (PEEP).

True reperfusion injury that directly adversely impacts the clinical course of the patient occurs in ~10% of patients. In this condition, a capillary leak occurs in (and is limited to) the endarterectomized areas of the lung. The cause of the leak can be multifactorial. In its most dramatic form, it occurs within a few hours after operation and is associated with profound desaturation. Edema-like fluid, sometimes with a bloody tinge, is suctioned from the endotracheal tube.

Once the capillary leak has been established, treatment is supportive, since 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 is diuresed aggressively, even if this requires ultrafiltration. The patient's ventilatory status may be dramatically position sensitive. The FiO_2 is kept as low as is compatible with an oxygen saturation of 90%. A careful titration of PEEP is performed, 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 ppm can improve gas exchange.

Frank blood from the endotracheal tube, which fortunately is very rarely seen, generally signifies a mechanical violation of the blood airway barrier that has occurred at surgery, usually unrecognized at the time. It is also occasionally seen when a patient preoperatively has a necrotic cavity of the lung (this is rare because of the bronchial arterial supply, as discussed earlier) which is revascularized by endarterectomy of the feeding vessels, previously blocked. Airway bleeding 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.

Residual pulmonary hypertension may result in right heart failure with hemodynamic instability, and sometimes also hypoxia, while reperfusion injury of the lung results in hypoxia. If these conditions are life-threatening, and considered reversible given adequate support and time, extracorporeal membrane oxygenation (ECMO) support may be initiated. Venoarterial support is used for a hemodynamic problem; removal of venous blood unloads the right ventricle and pulmonary artery, and the circuit provides gas exchange and increased cardiac output as blood is returned to the arterial side. If the issue is only the lack of oxygenation, then venovenous ECMO is used.

Results

The UCSD has the world's largest experience in this operation, now exceeding 3,700 patients. The mortality rate is in the range of 1–2%, with the vast majority of patients returned to normal activity.⁶

The mean patient age in the last 2,500 patients was 52 years, with a range of 14 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, or coronary artery bypass grafting.

The reduction in pulmonary pressures and resistance to normal levels and a corresponding improvement in pulmonary blood flow and cardiac output are generally immediate and, in general, these changes can be assumed to be permanent. With the elimination of the right ventricular pressure overload, the right ventricular geometry reverts to normal, and right atrial and right ventricular enlargement regresses.

At 1 year after the operation, 95% of patients remain in NYHA functional Class I or II.

With our increasing experience and many referrals, we are encountering more patients with Type III disease, and also accepting 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.

Conclusion

Pulmonary hypertension caused by chronic pulmonary embolism is a condition which is under-recognized, and carries a poor prognosis. Medical therapy is ineffective in prolonging life and only transiently improves the symptoms.

Pulmonary endarterectomy is technically demanding, and requires careful dissection of the pulmonary artery planes and the use of circulatory arrest. There is a distinct learning curve for the procedure. The postoperative management is more complex than in the usual heart surgery case. However, excellent short- and long-term results can be achieved. The mortality for thromboendarterectomy at our institution is now in the range of 1-2%, with sustained benefit.

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.

References

- 1 Lang IM. Chronic thromboembolic pulmonary hypertension—not so rare after all. *N Engl J Med* 2004;350(22):2236–2238
- 2 Lang IM, Madani M. Update on chronic thromboembolic pulmonary hypertension. *Circulation* 2014;130(06):508–518
- 3 Jamieson SW, Kapelanski DP. Pulmonary endarterectomy. *Curr Probl Surg* 2000;37(03):165–252
- 4 Nijkeuter M, Hovens MM, Davidson BL, Huisman MV. Resolution of thromboemboli in patients with acute pulmonary embolism: a systematic review. *Chest* 2006;129(01):192–197
- 5 Jamieson SW, Kapelanski DP, Sakakibara N, et al. Pulmonary endarterectomy: experience and lessons learned in 1,500 cases. *Ann Thorac Surg* 2003;76(05):1457–1462, discussion 1462–1464
- 6 Madani MM, Auger WR, Pretorius V, et al. Pulmonary endarterectomy: recent changes in a single institution's experience of more than 2,700 patients. *Ann Thorac Surg* 2012;94(01):97–103, discussion 103