The Royal College of Surgeons of England

Ann R Coll Surg Engl 2001; **83**: 427–430

Original article

Pulmonary thromboendarterectomy

Heyman Luckraz, John Dunning

Transplant Unit, Papworth Hospital, Cambridge, UK

Pulmonary thromboendarterectomy (PTE) provides a curative alternative to the otherwise fatal condition of chronic thromboembolic pulmonary hypertension (CTEPH). However, the condition is under-diagnosed due to a lack of awareness. An acceptable operative mortality of around 10% and long-term survival exceeding medical therapy or transplantation makes PTE a favourable choice for the treatment of CTEPH. Outcome is further optimised if the disease is diagnosed early and patients referred to specialised centres. An increase in the number of surgical procedures will also contribute to lower the mortality associated with this condition.

Key words: Pulmonary thromboendarterectomy – Chronic thromboembolic pulmonary hypertension – Early diagnosis

Chronic thromboembolic pulmonary hypertension (CTEPH) is an under-recognised, under-diagnosed, progressive and fatal condition.[1] It stems from the incomplete resolution of, or recurrent, pulmonary thromboembolic episodes. Currently, it is managed either palliatively with vasodilators and anticoagulants or surgically by transplantation or pulmonary thromboendarterectomy (PTE). Although Hollister and Cull[2] reported the first PTE in 1956, only around 1000 such procedures have been carried out world-wide. In the UK, around 50 such procedures have been performed at Papworth Hospital over the last 3 years.[3]

The vast majority of CTEPH patients has idiopathic thromboembolism as their primary pathology although 5–10% of patients have a defined coagulation problem such as lupus anticoagulant, protein C deficiency, antithrombin III deficiency and heparin-induced antibody.[4] The prevalence of the disease is not known, but it is believed that it is not a rare disorder. This is based on postmortem studies, which showed that up to 1% necropsies had evidence of chronic massive thrombosis of the major pulmonary arteries.[5] Moreover, it is estimated that 1–5% of patients who have a pulmonary embolic (PE) episode will develop CTEPH.[6] In the US, over 500,000 symptomatic PE episodes have been reported annually.[7] This is still an under-estimate as necropsy studies show evidence of acute pulmonary clots in 30% of cases.[8]

In terms of treatment modality for CTEPH, medical therapy is seldom effective while survival after transplantation is associated with significant morbidity related to acute rejection and infection, and limited long-term results due to chronic rejection. PTE offers a better alternative to the grim outlook for these patients.

Correspondence to: Mr Heyman Luckraz, Transplant Specialist Registrar, Papworth Hospital, Papworth Everard, Cambridgeshire CB3 8RE, UK. Tel: +44 1480 830541; Fax: + 44 1480 364610; E-mail: Heyman.Luckraz@papworth-tr.anglox.nhs.uk

Untreated, the 5-year survival for CTEPH patients whose mean pulmonary artery pressure exceeds 50 mmHg, is only 10%.[1] Survival post-PTE is reported to be 90% and 81% at 1 and 3 years, respectively,[4] with an operative mortality of 7%.[9,10] Furthermore, over 95% of operated patients improve clinically as per the reported improved NYHA classification.[10]

As already mentioned, an estimated 5%[6] of patients who suffer an acute PE will survive to develop CTEPH. This is the result of incomplete resolution of the embolic material, which becomes organised and occludes or narrows the pulmonary arterial tree, leading to pulmonary hypertension. Four types are recognised as suggested by the Jamieson classification.[4,11]:

Type 1 – classical thrombus in the proximal artery (20%).

Type 2 – no visible thrombus present on initial inspection of the pulmonary artery (PA). The endarterectomy is still performed with the plane raised at mid-to-distal main PA or lobar arteries and carried distally (60%).

Type 3 – no visible thrombus proximally but there is distal intimal thickening (15%).

Type 4 – pulmonary hypertension not due CTEPH and thus inoperable (4A, Eisenmenger's syndrome; 4B, primary pulmonary hypertension).

Recanalisation of thrombus ensues but provides inadequate flow through the PA tree causing pulmonary hypertension (PHT). This leads to right heart strain which progresses to right ventricular hypertrophy and dilatation over time. Right heart failure ensues and death is an inevitable outcome if the PHT is not treated.[12]

Pre-operative assessment

The clinical presentation and physical examination of CTEPH patients are non-specific. The most common complaint is progressive exercise intolerance. The latter is usually investigated, but only in rare instances is PHT due to chronic thromboembolism considered. This is due to a lack of awareness for this potentially curable condition. Other symptoms include cough, syncope, palpitations and haemoptysis. A past history of deep vein thrombosis or pulmonary embolism is elicited in less than half of the patients.[13] The typical features of PHT and right heart failure are found on physical examination including wide-fixed splitting of the second heart sound and pulmonary flow murmurs.

Prothrombotic conditions are investigated through a thrombophilia screen. A lung perfusion/ventilation scan to detect major perfusion defects is performed if CTEPH is considered. The cardiac function and dimensions are estimated with transthoracic echocardiography. Tricuspid regurgitation is a common finding and is a result of altered right ventricular dimension secondary to PHT. Right heart catheterisation provides useful information of the right-sided cardiac pressures while the pulmonary haemodynamics are assessed with a pulmonary artery flotation catheter. The pulmonary tree is visualised at angiography using selective power injections of non-ionic contrast medium, which is well tolerated in these haemodynamically rather unstable patients. The PA anatomy can also be assessed with spiral-CT angiography, which confirms the mosaic perfusion characterising this pathology. On rare occasions, pulmonary angioscopy is necessary if the cause of the PHT is still unclear after the above-mentioned investigations.

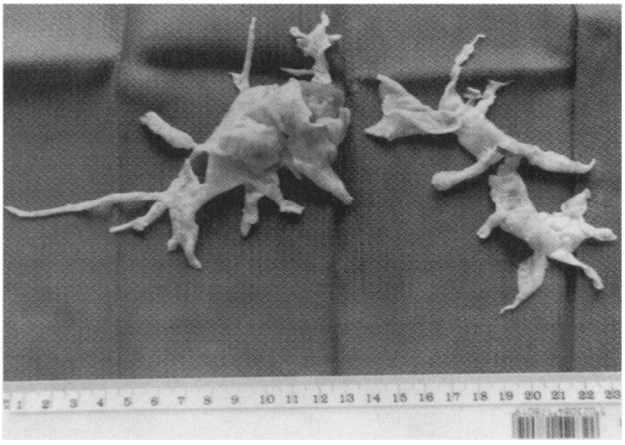

Figure 1 Specimen of endarterectomised material from the pulmonary arteries.

Surgical procedure

If CTEPH is confirmed, patients are referred for PTE. An inferior vena cava filter is inserted pre-operatively in all patients. Median sternotomy approach provides the best exposure for this procedure. The pericardial sac is opened and cardiopulmonary bypass (CPB) is instituted via bicaval venous cannulation for drainage and ascending aortic cannulation for oxygenated blood return. While the patient is being cooled to 18°C, the superior vena cava is mobilised to improve access to the right PA. Care is taken to avoid injury to phrenic nerves. A left ventricular vent is used to prevent cardiac distension when the heart fibrillates following the lowering of the core body temperature. A PA venting circuit is also used to protect the right ventricle for over-distension. Myocardial protection is achieved by using cold crystalloid cardioplegia and maintained by a

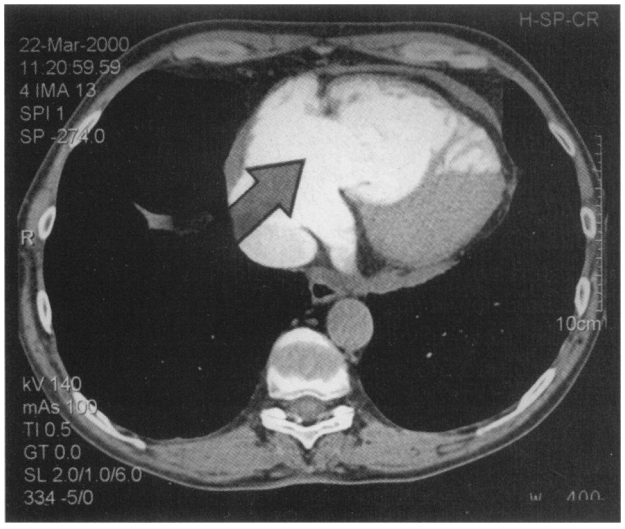

Figure 2 CT scan with contrast showing the enlarged right ventricle (arrow).

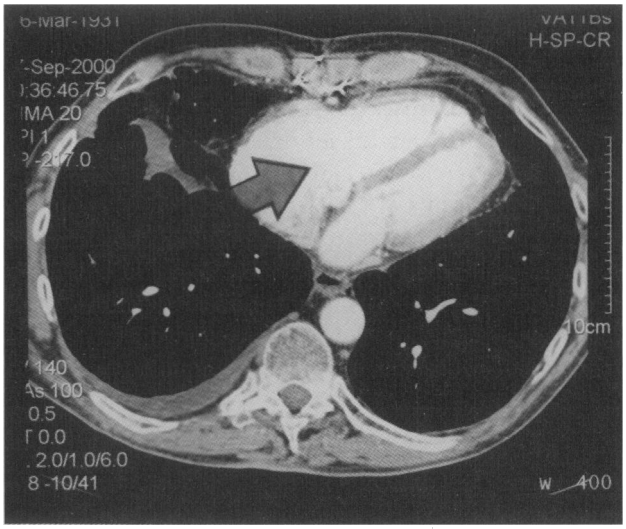

Figure 3 CT scan with contrast 3 months post PTE showing the remodelled right ventricular cavity.

cooling jacket, which is wrapped around he heart. Due to significant bronchial blood collateral flow, adequate exposure is achieved by intermittent periods of circulatory arrest with reperfusion. An endarterectomy plane is raised centrally in the pulmonary artery. The dissection must be carried distally to at least two divisions distal to the origin of the segmental arteries. The initial plane is created on the posterior wall of the PA and pursued circumferentially. Distal endarterectomy is achieved by a combination of traction, suction and gentle dissection using the Jamieson dissector. The procedure is carried out on both sides, as CTEPH is a bilateral disease. A typical specimen of the endarterectomised material is shown in Figure 1. Incomplete endarterectomy can be fatal as the right ventricle fails to cope with the persistent PHT. In this situation, patients cannot be weaned off the bypass circuit.

Once the endarterectomy is complete, CPB is resumed and the patient re-warmed. Additional cardiac procedures can be carried out during the re-warming phase and include closure of the secundum atrial septal defect (25% patients), coronary artery bypass grafting and valve replacement. Tricuspid regurgitation does not need attending to, as valvular competence is restored when the right ventricle remodels with time. On the other hand, pulmonary artery haemodynamics and pulmonary vascular resistance improve immediately after PTE. These changes are illustrated by one of our patients who was operated recently.

He was a 68-year-old man with 12-months history of progressive increase of shortness of breath and bed-bound on home oxygen. His relevant past medical history included manic depression on triple therapy (lithium, carbamezepine and largactil) and diabetes insipidus secondary to lithium. Haemodynamic measurements pre-operatively were pulmonary arterial pressure (PAP) of 110/45 mmHg (mean PAP = 55 mmHg) and PaO_2 (pulmonary artery) saturation of 48% on room air. Systemic blood pressure was 150/95 mmHg with O_2 saturation of 85% on room air. Cardiac index measured via a pulmonary artery flotation catheter, was 1.6 $l.min^{-1}.m^{-2}$. These parameters improved immediately post PTE and at 3 months postoperatively were PAP 30/10 mmHg (mean = 16 mmHg) and cardiac index of 2.65 $l.min^{-1}.m^{-2}$. The right ventricular dimensions, as assessed by transthoracic echocardiography and CT scans (Figs 2 & 3) revealed marked improvement. Moreover, he showed an improvement in his psychiatric condition requiring only carbamezepine and largactil as long-term medication. Thus, his diabetes insipidus also resolved.

Postoperative management

Although the surgical approach involved in the management of CTEPH through PTE requires meticulous skill, the success of CTEPH treatment also includes a short intense postoperative care.[14] Patients are ventilated for at least 48 h with PEEP as tolerated. Nitric oxide is sometimes used to overcome hypoxia secondary to shunting of blood away from the normal pulmonary areas. Reperfusion pulmonary oedema (RPE) has been described in up to 20% of patients being fatal in about 2%.[12] This abnormality may develop up to 72 h post PTE.[15] It is believed that RPE represents a pressure phenomenon in previously hypoperfused vasculature in

association with alterations in surfactant in lung areas distal to the occluded PA.[16] RPE is managed conservatively and usually resolves after a few days. Cerebral complications such as temporary change of affect and motor weakness are noted in 10% of postoperative patients and represent altered cerebral vascular autoregulation during the circulatory arrest periods. These symptoms usually improve shortly after. Occasionally, haemoptysis may develop as a result of over-zealous intrapulmonary dissection. All patients are anticoagulated with low molecular weight heparin initially and later with warfarin aiming for an INR of 2.5–3.5.

References

1. Riedel M, Stanek V, Widimsky J, Preovsky I. Long term follow up of patients with pulmonary thromboembolism. Late prognosis and evolution of haemodynamic and respiratory data. *Chest* 1982; **81**: 151–8.
2. Hollister LE, Cull VL. The syndrome of chronic thromboembolism of the major pulmonary arteries. *Am J Med* 1956; **21**: 312–20.
3. Dunning J. Personal communication.
4. Mo M, Kapelanski DP, Mitruka SN, Auger WR, Fedullo PF, Channick RN *et al.* Re-operative pulmonary thromboendarterectomy. *Ann Thorac Surg* 1999; **68**: 1770–7.
5. Presti B, Berthrong M, Sherwin RM. Chronic thrombosis of major pulmonary arteries. *Hum Pathol* 1990; **21**: 601–6.
6. Fedullo PF, Auger WR, Channick RN, Moser KM, Jamieson SW. Chronic thromboembolic pulmonary hypertension. *Clin Chest Med* 1995; **16**: 353–74.
7. Anderson Jr FA, Wheeler HB, Goldberg RJ, Hosmer DW, Patwardhan NA, Jovanovic B *et al.* A population based perspective of the hospital incidence and case fatality rates of deep vein thrombosis and pulmonary embolism. The Worcester DVT Study. *Arch Intern Med* 1991; **151**: 933–8.
8. Diebold F, Lohres U. Venous thrombosis and pulmonary embolism. *Pathol Res Pract* 1991; **187**: 260–6.
9. Kramm T, Mayer E, Dahm M, Guth S, Menzel T, Pitton M *et al.* Long term results after thromboendarterectomy for chronic pulmonary embolism. *Eur J Cardiothorac* 1999; **15**: 579–84.
10. Moser KM, Auger WR, Fedullo PF, Jamieson SW. Chronic thromboembolic pulmonary hypertension: clinical picture and surgical treatment. *Eur Resp J* 1992; **5**: 334–42.
11. Jamieson SW. Pulmonary thromboendarterectomy. In: Franco KL, Putman JD. (eds) *Advanced Therapy in Thoracic Surgery*. Hamilton: Decker, 1998, 310–8.
12. Dunning J, McNeil K. Pulmonary thromboendarterectomy for chronic thromboembolic pulmonary hypertension. *Thorax* 1999; **54**: 755–8.
13. Jamieson SW. Pulmonary thromboendarterectomy. *Heart* 1998; **78**: 118–20.
14. Wilson WC, Vuylsteke A. Anaesthesia for pulmonary thrombo-endarterectomy. In: Ghosh S, Latimer R. (eds) *Thoracic Anaesthesia: Principles and Practice*. London: Butterworth-Heinemann, 1999, 223–34.
15. Jamieson SW, Auger WR, Fedullo PF, Channick RN, Kriett JM, Tarazi RY *et al.* Experience and results with 150 pulmonary thromboendarterectomy operations over a 29-month period. *J Thorac Cardiovasc Surg* 1993; **106**: 116–27.
16. Louber GP, Cohen E, Neustein S. Pulmonary reperfusion oedema associated with pulmonary thromboendarterectomy. *J Cardiothorac Vasc Anaesth* 1998; **12**: 353–7.