



Original

## Chronic thromboembolic pulmonary hypertension: surgical treatment with thromboendarterectomy

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## ABSTRACT

**Background and objectives:** Pulmonary thromboendarterectomy (PTE) is considered the potential curative treatment for chronic thromboembolic pulmonary hypertension (CTEPH). We analysed the results of the PTE application in our institution.

**Patients and methods:** From February 1996 to December 2007, 30 patients with CTEPH underwent video-assisted PTE. Preoperative hemodynamic data were: systolic pulmonary artery pressure (SPAP) 87±17 mmHg, mean pulmonary artery pressure (MPAP) 51±11 mmHg, pulmonary total resistance 1067±485 dynes·s·cm<sup>-5</sup>, pulmonary vascular resistance 873±389 dynes·s·cm<sup>-5</sup> and cardiac index 2.2±0.5 l/min/m<sup>2</sup>. We analysed the influence of several factors on hospital mortality and survival, and we performed partial analysis of mortality since 2004.

**Results:** PTE resulted in significant improvements in SPAP ( $P<0.001$ ), MPAP ( $P=0.001$ ) and cardiac index ( $P<0.001$ ). Hospital mortality was 17% (5/30) (95% confidence interval, 6%-35%). From 2004, it dropped to 5% (1/20) (95% confidence interval, 0%-25%). Hospital mortality was influenced by preoperative pulmonary total resistance, preoperative pulmonary vascular resistance, postoperative SPAP, reduction of MPAP, reperfusion pulmonary oedema and residual postoperative pulmonary hypertension ( $P=0.036$ ;  $P=0.018$ ;  $P=0.013$ ;  $P=0.050$ ;  $P=0.050$ ;  $P=0.030$ ;  $P=0.045$ ). Survival after PTE, including hospital mortality, was 76±9% at 10 years. Through long-term follow-up, functional status ( $P=0.001$ ), 6 min walking distance ( $P=0.001$ ), end-diastolic right ventricle size ( $P<0.001$ ), and tricuspid regurgitation ( $P<0.001$ ) significantly improved.

**Conclusions:** PTE effectively reduces pulmonary hypertension and offers CTEPH patients a substantial improvement in survival and quality of life.

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### Hipertensión pulmonar tromboembólica crónica: tratamiento mediante tromboendarterectomía quirúrgica

## RESUMEN

**Introducción:** La tromboendarterectomía pulmonar (TP) constituye el tratamiento potencialmente curativo de la hipertensión pulmonar tromboembólica crónica (HTPTC). Analizamos los resultados de la aplicación de la TP en nuestra institución.

**Pacientes y métodos:** Entre febrero de 1996 y diciembre de 2007 se realizó TP videoasistida a 30 pacientes con HTPTC. Los datos hemodinámicos preoperatorios fueron (valores medios±desviación estándar): presión sistólica pulmonar (PSP), 87±17 mmHg; presión arterial pulmonar media (PAPm), 51 ± 11 mmHg; resistencia pulmonar total, 1.067 ± 485 dinas·s·cm<sup>-5</sup>; resistencia vascular pulmonar, 873 ± 389 dinas·s·cm<sup>-5</sup>, e índice cardíaco, 2,2 ± 0,5 l/min/m<sup>2</sup>. Se han analizado los factores que influyeron en la mortalidad hospitalaria y la supervivencia, además de realizarse un análisis parcial de la mortalidad a partir de 2004.

## Palabras clave:

Hipertensión pulmonar tromboembólica crónica  
Tromboendarterectomía pulmonar  
Tromboembolia pulmonar

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**Resultados:** Tras la TP se objetivó un descenso tanto de la PSP ( $p < 0,001$ ) como de la PAPm ( $P = 0,001$ ) y un aumento del índice cardíaco ( $p < 0,001$ ). La mortalidad hospitalaria registrada fue del 17% (5/30; intervalo de confianza del 95%, 6–35%); a partir de 2004 se redujo al 5% (1/20; intervalo de confianza del 95%, 0–25%). La resistencia pulmonar total y la resistencia vascular pulmonar preoperatorias, la PSP postoperatoria, el descenso porcentual de la PSP y de la PAPm, la presencia de edema de reperfusión y la persistencia de la HTP evidenciaron asociación con la mortalidad hospitalaria ( $p = 0,036$ ;  $p = 0,018$ ;  $p = 0,013$ ;  $p = 0,050$ ;  $p = 0,050$ ;  $p = 0,030$ ;  $p = 0,045$ , respectivamente). La supervivencia actuarial a 10 años, incluyendo la mortalidad hospitalaria, fue del  $76 \pm 9\%$ . Durante el seguimiento mejoró la clase funcional ( $p = 0,001$ ), aumentó la distancia recorrida en la prueba de la marcha de 6 min ( $p = 0,001$ ) y se redujeron tanto el diámetro telediastólico del ventrículo derecho ( $p < 0,001$ ) como el grado de regurgitación tricuspídea ( $p < 0,001$ ).

**Conclusiones:** La TP mejora la hemodinámica pulmonar, prolonga la supervivencia y optimiza el estado funcional de pacientes con HTPC.

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## Introduction

Chronic pulmonary thromboembolism (CPTE) and chronic thromboembolic pulmonary hypertension (CTEPH) are aetiopathogenically linked entities that independently lead to high morbidity and mortality rates. In 1–5% of cases of CPTE, the fibrinolytic system fails and the thrombus organises itself and integrates the wall of the pulmonary arterial system, causing CTEPH independently of the recurrence of chronic thromboembolic phenomena.<sup>1,2</sup> Although the development of CTEPH has an aetiological link with the prior presence of CPTE, more than 50% of patients diagnosed with CTEPH do not present antecedents of clinically documented CPTE, and therefore it is probable that the incidence of this disease is greater than that which may be estimated based on CPTE.<sup>3</sup> The fundamental symptomatology of CTEPH is non-specific (dyspnoea upon exertion and fatigue) and the progressive development of pulmonary hypertension (PH) can cause the development of compensatory right ventricular hypertrophy. Once it has been established, PH develops progressively; determining factors in its progression are the slow thrombosis of the rest of the vascular tree secondary to stasis and reactive histopathological changes in vascular territories not initially affected.

The underdiagnosis of CTEPH is relevant since it is possible to cure patients affected by this type of PH via pulmonary thromboendarterectomy (PTE).<sup>3–7</sup> PTE is a surgical procedure which is technically very demanding and is carried out in a very small number of centres throughout the world.<sup>8,9</sup> This article presents experience in the application of PTE as a method of treating CTEPH in this institution and analyses the results in terms of the procedure's mortality rate and its efficacy with regard to the regression of PH, improvement in the functional status and long-term survival.

## Patients and methods

### Population and criteria for selecting surgical patients

The series includes 30 consecutive patients affected by CTEPH on whom PTE was performed between February 1996 and December 2007. Their mean age ( $\pm$  standard deviation) was  $54 \pm 12$  and 63% (19/30) were male. Inclusion criteria were the following: a) symptomatic proximal CTEPH; b) mean pulmonary artery pressure (mPAP) greater than 30mmHg not responding to pharmacological treatment; and c) pulmonary vascular resistance (PVR) greater than 300 dynes  $s \cdot cm^{-5}$ . The condition of proximal thromboembolic pulmonary disease (disease of the main pulmonary, lobular or proximal segmental arteries) was a fundamental criteria for selecting patients for surgical treatment, since the determining factor for resectability is the localisation and spread of obstructions. Exclusion criteria included the concomitant presence of obstructive or seriously

restrictive pulmonary disease, malign neoplasm, terminal non-cardiac disease and severe left ventricular dysfunction.

### Diagnostic assessment: general protocol for pulmonary hypertension and specific protocol for chronic thromboembolic pulmonary hypertension

Initially, the general PH protocol was applied to patients: clinical examination, chest x-ray, electrocardiogram, echocardiogram, respiratory function tests and scintigraphy of pulmonary perfusion. Doppler echocardiogram confirmed the diagnosis of PH by estimating systolic pulmonary artery pressure (SPAP), and scintigraphy of pulmonary perfusion was the test used to distinguish patients in which PH could be of thromboembolic aetiology: a normal scintigraphy excludes the diagnosis of PH due to pulmonary embolism.<sup>3,7</sup> The specific diagnosis protocol was applied to patients with segmental perfusion defects to confirm the presence of chronic pulmonary disease: chest scan and pulmonary arteriography. Pulmonary arteriography, the reference test for confirming the diagnosis of PH of thromboembolic origins, is essential in determining the viability of PTE in terms of localisation, the size and spread of the thromboembolic disease.<sup>3,9</sup> However, the findings of the helical chest scan with intravenous contrast showed an excellent correlation with the postoperative anatomopathological prognostic classification, which linked the localisation and spread of the thrombus with the result of PTE.<sup>9,10</sup> Cardiac catheterisation was performed during the same procedure as the pulmonary arteriography, which confirmed the diagnosis of PH by invasive means, as well as assessing the severity of the PH and reactivity of the pulmonary tree.

Coronary catheterisation was performed on patients undergoing PTE who were over 45 or presented risk factors for coronary artery disease (antecedents of ischaemic heart disease based on clinical data and/or non-invasive tests, left ventricular dysfunction and the presence of one or more cardiovascular risk factors).

The degree of left ventricular dysfunction and the severity of tricuspid regurgitation were assessed via Doppler echocardiography. Left ventricular dysfunction was assessed based on dilatation of the right ventricle (mild: 29–33 mm; moderate: 34–39 mm; severe > 39mm) and fractional change in the area of the right ventricle (mild: 25–31%; moderate: 18–24%; severe  $\leq$  17%). Tricuspid regurgitation was classified from the regurgitating area (class I < 4 cm<sup>2</sup>; class II: 4–6 cm<sup>2</sup>; class III: 6–8 cm<sup>2</sup>; class IV > 8 cm<sup>2</sup>).

Following confirmation of the thromboembolic origin of PH, patients underwent an exhaustive study of thrombophilia via functional, antigen and molecular biology tests: determination of resistance to activated protein C, functional determination of antithrombin III, protein C, protein S, lupus anticoagulant and anticardiolipin antibodies of IgG and/or IgM isotype, homocysteine, plasminogen, prothrombin gene mutation 20210, the factor V Leiden gene and the MTHFR gene.

### Functional assessment

The subjective assessment of functional status was performed according to adaptation of the functional classification of the New York Heart Association by the World Health Organisation (WHO) for PH. The 6 min walking test was used as an objective test to assess functional status.

### Characterisation of chronic thromboembolic pulmonary disease

Chronic thromboembolic pulmonary disease was characterised according to the anatomopathological prognostic classification that links the localisation and spread of the thrombus with the result of PTE.<sup>10</sup> This classification established the existence of four anatomopathological types of CTEPH: type 1 (20% of cases), consisting of thrombus that occupy the main pulmonary arteries and are revealed immediately after performing a pulmonary arteriotomy; type 2 (70% of cases), consisting of intimal thickening and fibrosis proximal to the segmental arteries, with no thrombus in the main pulmonary arteries; type 3 (10% of cases), consisting of thromboembolic disease in the distal segmental and subsegmental arteries only, and type 4, consisting of distal arteriolar vasculopathy without macroscopic thromboembolic disease, which cannot be operated on and presents due to a discrepancy between the degree of objective macroscopic obstruction and the associated haemodynamic alteration.<sup>2,11</sup>

### Surgical technique

PTE was performed according to the protocol created by the group from the University of California (San Diego, US).<sup>12,13</sup> The surgical technique consisted of bilateral pulmonary endarterectomy via median sternotomy with cardiopulmonary bypass and intermittent periods of circulatory arrest and deep hypothermia. In the majority of patients both pulmonary arteries are affected and therefore bilateral PTE must be performed.<sup>14</sup> The bronchial circulation of patients with CTEPH is extraordinarily developed and therefore, in order to define the correct endarterectomy plane, the pulmonary arteries must be maintained exsanguis during periods of circulatory arrest.<sup>12,13</sup> Periods of circulatory arrest with deep hypothermia were established at 10 min, alternated with 5 min of reperfusion, completing the PTE with an accumulated time of circulatory arrest in each pulmonary artery of under 20 min in the majority of cases.

Access and view of the most distal localisations of the thromboembolic disease was optimised with the use of a rigid 5mm Hopkins II hysteroscope (Karl Storz, Tuttlingen, Germany) with optics of 0°-30°-45° connected to a Twinvideo video camera (Karl Storz, Tuttlingen, Germany).

The comprehensive treatment of the chronic thromboembolic disease was completed with the insertion of a Greenfield filter into the inferior vena cava and the establishment of a permanent anticoagulation regimen.<sup>13</sup>

### Efficacy criteria for pulmonary thromboendarterectomy

The efficacy of PTE was assessed via the reduction in figures relating to pulmonary artery pressure. A reduction greater than 50% of the basal value for SPAP and mPAP was considered clinically significant.

### Persistence of pulmonary hypertension and reperfusion oedema

Specific complications of PTE are the persistence of PH and reperfusion pulmonary oedema. The persistence of PH was defined as a reduction in mPAP of less than 50% of the preoperative value, and reperfusion oedema as the development of hypoxaemia and radiological infiltrates in areas of reperfused endarterectomy.<sup>15</sup>

Significant reperfusion oedema was defined as that which required prolonged mechanical ventilation (> 24 hr) or reintubation and reconnection of mechanical ventilation.

### Follow-up

Postoperative follow-up was performed via a consultation in the specialist PH reference unit within this institution.<sup>9</sup> Follow-up began on the date of the surgical intervention. Visits took place every three months during the first year and subsequently every six months, with 100% compliance. The follow-up protocol included the performance of a Doppler transthoracic echocardiography and the 6 min walking test. The echocardiogram was used to estimate residual SPAP and analyse the geometric redevelopment and functional recovery of the right ventricle following PTE.<sup>9</sup> The 6 min walking test was used to objectively assess improvements in patients functional capacity following surgical intervention.

### Statistical analysis

The influence of clinical, demographic, operative and haemodynamic variables on hospital mortality and survival were analysed and a partial analysis of hospital mortality since 2004 was performed. Continuous variables were mean  $\pm$  standard deviation and categorical variables frequencies. The association between categorical variables was analysed via Pearson's  $\chi^2$  test or Fisher's exact test. Quantitative variables were analysed by applying Student's t-test or the Mann-Whitney U test based on the normality of distributions according to the Shapiro-Wilk test.

Survival curves were calculated using the Kaplan-Meier method, and the logrank test was used to compare survival between the groups based on each of the prognostic factors considered.

A value of  $p < 0.05$  was considered statistically significant.

## Results

### Preoperative functional status

A total of 93% of patients (28/30) were in functional class III-IV of the WHO classification. The distance covered during the 6 min walking test was  $340 \pm 124$ m. A total of 63% presented criteria of cardiac insufficiency, and in the immediate preoperative period, 27% required inotropic support and 59% epoprostenol.

### Preoperative haemodynamics and right ventricular function

Preoperative SPAP was  $87 \pm 17$ mmHg (range: 53–119) and mPAP  $51 \pm 11$ mmHg (range: 31–67). Total pulmonary resistance was  $1.067 \pm 485$  dynes-s-cm<sup>75</sup> and PVR  $873 \pm 389$  dynes-s-cm<sup>75</sup>. Cardiac index was  $2.2 \pm 0.5$ L/min/m<sup>2</sup>.

A total of 86% of patients presented right ventricular dysfunction, which was severe in 83%. The telediastolic diameter of the right ventricle, viewed with echocardiography, was  $48.9 \pm 5.4$ mm. A total of 93% of the series presented tricuspid regurgitation, which was severe in 40%.

### Chronic pulmonary thromboembolism and thrombophilia

A total of 90% of patients presented antecedents of CPE and the presence of hypercoagulability disorder was found in 60%, with the predominance of lupus anticoagulant.

### Intraoperative pathologic anatomy

A total of 93% (28/30) presented proximal chronic pulmonary disease (types 1 and 2); of the other two types, type 3 predominated



(figs. 1 and 2). PTE was performed with aortic crossclamping for  $132 \pm 31$  min, and a total circulatory arrest time of  $38 \pm 16$  min was required.

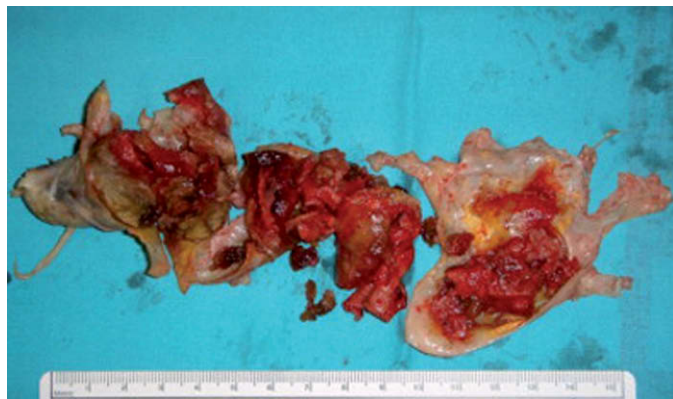
#### Postoperative haemodynamics

Haemodynamic evolution immediately following PTE was satisfactory (fig. 3). There was a significant reduction in SPAP ( $87 \pm 17$  compared with  $56 \pm 20$  mmHg;  $p < 0.001$ ) and mPAP ( $51 \pm 11$  compared with  $30 \pm 10$  mmHg;  $p = 0.001$ ) in absolute terms, and an increase in cardiac index ( $2.3 \pm 0.5$  compared with  $3.3 \pm 0.6$  L/min/m<sup>2</sup>;  $p < 0.001$ ). A clinically significant reduction in SPAP was obtained in 41% of patients and mPAP in 44%.

#### Hospital morbidity and mortality

A total of 43% of patients (13/30) developed reperfusion oedema and 40% (12/30) significant reperfusion oedema.

The hospital mortality recorded was 17% (5/30; confidence interval 95%, 6–35%). The cause of death was respiratory failure in 80% (4/5) and multiorgan failure in the remaining patients. The partial analysis of mortality from 2004 showed that hospital mortality dropped by 5% (1/20; confidence interval 95%, 0–25%). The persistence of PH ( $p = 0.045$ ), the presence of reperfusion oedema ( $p = 0.030$ ) and



**Figure 1.** Surgical specimen of type 1 chronic thromboembolic pulmonary disease.



**Figure 2.** Surgical specimen of type 2 chronic thromboembolic pulmonary disease.

significant reperfusion oedema ( $p = 0.021$ ) showed an association with hospital mortality. Among the preoperative haemodynamic variables, total pulmonary resistance ( $1,576 \pm 547$  compared with  $940 \pm 395$  dynes·s·cm<sup>-5</sup>;  $p = 0.036$ ) and PVR ( $1,389 \pm 438$  compared with  $744 \pm 259$  dynes·s·cm<sup>-5</sup>;  $p = 0.018$ ) showed an association with hospital mortality. Among the postoperative haemodynamic variables, SPAP ( $80 \pm 25$  compared with  $51 \pm 15$  mmHg;  $p = 0.013$ ) and percentage drops in SPAP ( $11 \pm 30$  compared with  $38 \pm 23$ ;  $p = 0.050$ ) and mPAP ( $-9 \pm 12$  compared with  $39 \pm 22$ ;  $p = 0.050$ ) following PTE showed an association with hospital mortality.

#### Follow-up

The mean follow-up of the series was  $40 \pm 34$  months (range: 5–129). The actuarial survival at 10 years, including hospital mortality, is  $76 \pm 9\%$  (fig. 4). If hospital mortality is excluded, actuarial survival at 10 years for patients who underwent PTE is  $92 \pm 8\%$ . Reperfusion oedema ( $p = 0.005$ ) and the persistence of PH ( $p = 0.029$ ) behaved as factors for a poor prognosis for long-term survival.

The distance covered in the 6 min walking test following PTE was  $472 \pm 78$  m, which was significantly greater than the preoperative distance ( $p = 0.001$ ). With respect to the subjective assessment of functional status, during follow-up 90% of patients presented WHO functional classes I–II, and there were no cases of class IV. This represented a statistically significant improvement on the preoperative functional status, in which 93% of the series were placed in WHO functional classes III–IV ( $p = 0.001$ ).

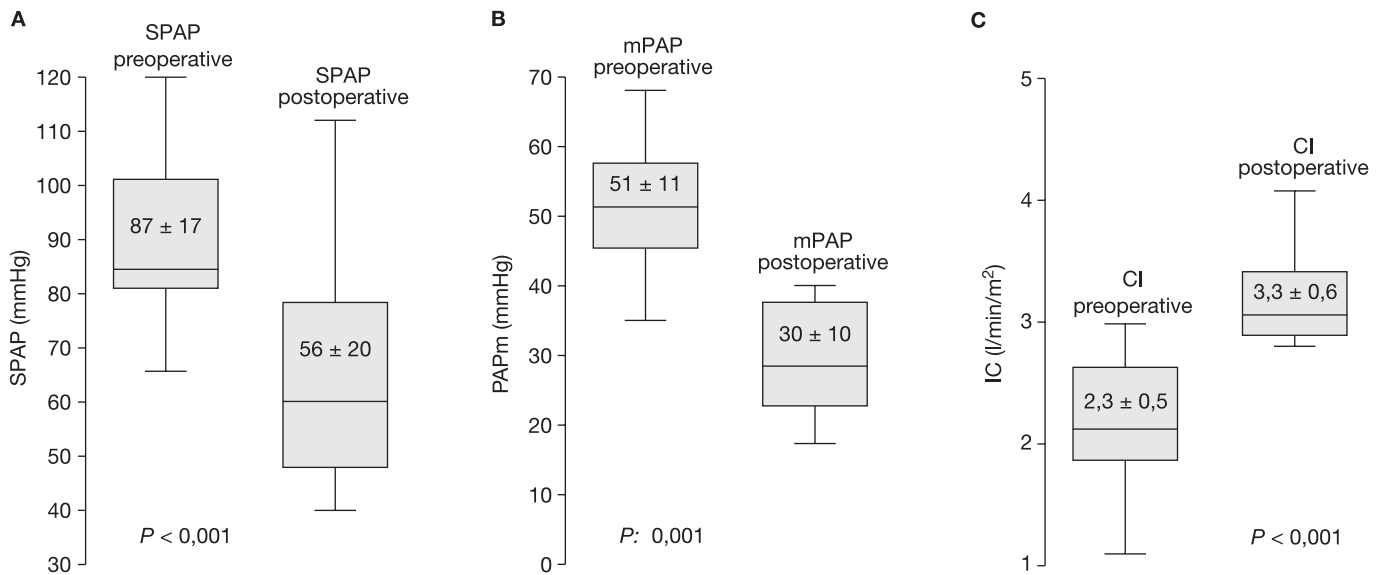
Following PTE, 80% of patients presented tricuspid regurgitation classes 0–I, and the remainder class II. This distribution of the postoperative tricuspid regurgitation represents a significant reduction with respect to that observed prior to the intervention, when 70% of cases presented tricuspid regurgitation classes III–IV ( $p < 0.001$ ). Another marker for the optimisation of right ventricular function is the significant reduction in the telediastolic diameter of the right ventricle to  $34 \pm 6$  mm ( $p < 0.001$ ).

The echocardiographic estimation of SPAP during objective follow-up maintained the significant drop in SPAP following the pulmonary endarterectomy in the long term ( $p < 0.001$ ).

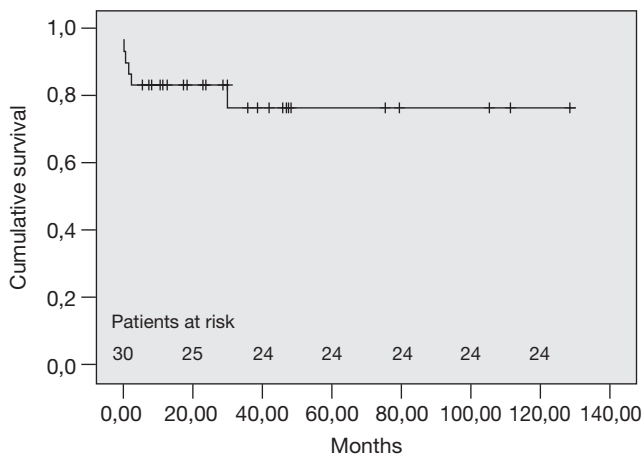
#### Discussion

CTEPH is an underdiagnosed fatal disease; the majority of patients die due to right ventricle failure.<sup>2,4,7,16</sup> Given the obstructive nature of CTEPH, medical treatment is not effective in prolonging survival and only improves the clinical picture temporarily.<sup>8,17</sup> Without surgical treatment, survival is very low and is associated with PH type at the time of the diagnosis.<sup>18</sup> PTE has been established as the treatment of choice for curing CTEPH.<sup>3,4,7,19,20</sup> The only alternative to PTE is a lung transplant although this must be considered as second line treatment, since PTE presents lower hospital mortality and greater long-term survival, without the inconvenients of chronic immunosuppression and chronic rejection of the new lung.<sup>5,9,17,21</sup>

The selection of suitable patients is a determining factor in the success of the surgical intervention.<sup>7</sup> In patients with proximal chronic pulmonary disease (types 1 and 2), PTE obtains excellent results in terms of the reduction in pulmonary artery pressure and PVR, with hospital mortality lower than 3%.<sup>3</sup> This series mainly consists of patients with proximal CTEPH, although there is a marginal percentage of patients operated on in which the proximal disease coexisted with a certain degree of distal disease, and in which it was supposed that the PTE significantly, but not wholly, contributed to the resolution of PH; the surgical risk of such patients is significantly greater, since the distal reactive disease more frequently leads to reperfusion oedema and the persistence of PH following PTE. In this respect, when the extent of the PH does not have a direct relationship with the degree of obstruction, it must be



**Figure 3.** Haemodynamic evolution immediately following pulmonary thromboendarterectomy is satisfactory, with a statistically significant reduction in systolic pulmonary artery pressure (A) and mean pulmonary artery pressure (mPAP) and a statistically significant increase in cardiac index (C). CI: Cardiac index; mPAP: mean pulmonary artery pressure; SPAP: systolic pulmonary artery pressure.



**Figure 4.** Survival of the series following the pulmonary thromboendarterectomy.

considered that the reactive arteriopathic changes in the vascular tree, which are not initially affected by the vascular obstruction but subjected to excess pressure and flow, are determining factors in the progression of PH.<sup>13</sup> In these patients, PTE yields few benefits and hospital mortality is greater than 25%.<sup>8,11</sup> PTE must be indicated as a soon as possible once a diagnosis of CTEPH has been established to prevent the development of type 4 disease from limiting or contraindicating the curative potential of the surgical intervention.<sup>19,20</sup>

In terms of the immediate haemodynamic development of the patient following the PTE, SPAP and mPAP showed a statistically significant drop in absolute values. The fact that less than half of the series initially obtained clinically significant reduction in SPAP and mPAP (reduction greater than 50% of the base value) is in keeping with the findings of other work groups<sup>23,24</sup> who have shown the presence of vascular hypertension associated with immediate postoperative stress which, once resolved, facilitates the relaxation of the reactive vasculature of the small vessel and, therefore, figures for pulmonary artery pressure.

In terms of hospital mortality, the comparative reference is 5% according to the group from the University of California (San Diego, US), which has experience of more than 2000 PTEs.<sup>7,12</sup> The hospital mortality recorded in this series was 17% (5/30), which is close to the 10-15% recorded by other groups with similar experience.<sup>4,25-28</sup> The partial analysis of hospital mortality from 2004, period in which 2/3 of the series were operated on (n = 20) showed that mortality had dropped by 5% (1/20). This partial analysis of mortality, also performed by other work groups,<sup>29</sup> establishes hospital mortality after obtaining determining factors for its reduction.<sup>9,30</sup> a) optimising the diagnostic assessment and selection of patients receiving surgical treatment; b) increasing the experience of the surgical team and c) implementing advances in the handling of postoperative complications specific to this procedure.

Among the haemodynamic variables, an association was obtained with hospital mortality, SPAP figures, total pulmonary resistance and preoperative PVR, and percentage reduction in SPAP and mPAP following PTE. Although these results must be interpreted with reserve, since they are obtained from an univariate analysis, they do confirm the findings of other work groups.<sup>4,20</sup>

The actuarial survival of patients surviving PTE is excellent: 92 ± 8% at 10 years. In this series, the persistence of PH behaved as a determining factor in survival. This is in keeping with that communicated by the majority of authors, who report that the determining factor of the vital prognosis and clinical improvement following the intervention is the degree to which resistance and pulmonary artery pressure has reduced.<sup>4,20</sup>

On the other hand, PTE is not only effective in terms of significantly reducing and maintaining the figures for pulmonary artery pressure, rather it also achieves functional recovery of the right ventricle.<sup>15,16,20</sup> In this series, the figures for SPAP were maintained in the long term and geometric remodelling of the right ventricle was noted from 2 markers for ventricular function and dimension: a) significant reduction in the degree of tricuspid regurgitation from 0-1 in 80% of patients and b) significant reduction in the telediastolic diameter of the right ventricle.

Another aspect facilitating the assessment of the efficacy of PTE is the improvement in patients' functional status following the intervention.<sup>3</sup> In this series, during follow-up, 90% of patients

presented WHO functional class I-II, while before the intervention more than 90% were placed in functional class III-IV. Likewise, there was also a significant increase in the distance covered in the 6 min walking test.

In short, CTEPH is an underdiagnosed fatal disease; medical treatment only improves symptomatology temporarily. PTE is the treatment of choice for anatomically favourable cases. The only therapeutic alternative to PTE is a lung transplant, which yields worse results in terms of survival. Obtaining satisfactory haemodynamic results following PTE is associated with vital prognosis and improvement of both the functional status and patients' quality of life. Direct interaction between the different members of a multidisciplinary group (pulmonologists, cardiologists, radiologists, anaesthesiologists, intensive medicine specialists and heart surgeons) and the selection of anatomically favourable cases are essential for optimising the results of PTE.

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