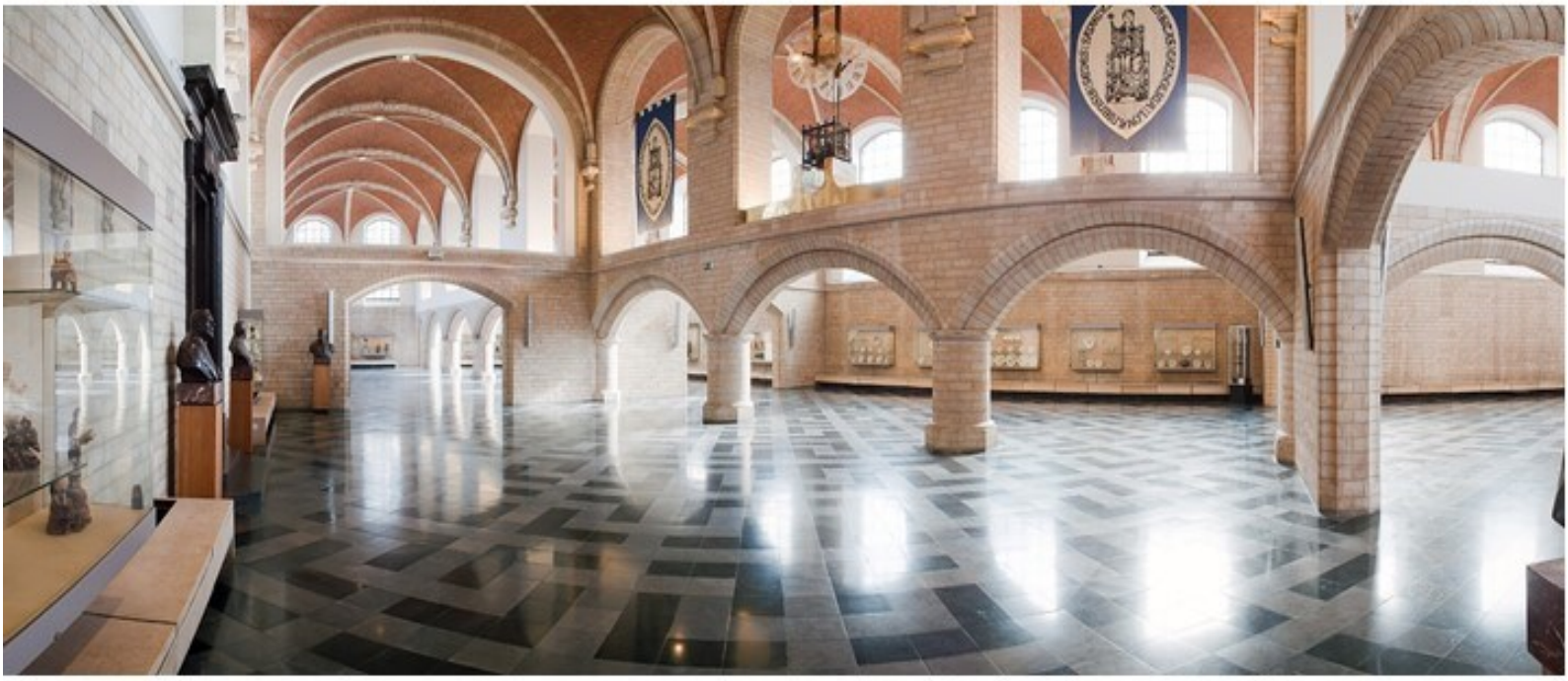
ICA International CTEPH Association

INTERNATIONAL CTEPH CONFERENCE 2017



Abstract Book

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Aula Pieter De Somer, Leuven, Belgium

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Pathophysiology, epidemiology, diagnosis and prognosis

ICC17-1. PULMONARY ARTERY SARCOMA AS A MIMIC CTEPH

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Introduction: Mechanical obstruction of pulmonary vessels leading to pulmonary hypertension is typically caused by organized thrombi remaining after pulmonary embolism. In rare cases, chronic thromboembolic pulmonary hypertension can be mimicked by other non-thrombotic diseases such as vasculitis, hydatidosis or tumor. In our case report, we describe rare intimal sarcoma mimicking CTEPH.

Case report: 65-year-old man with history of pulmonary embolism in March 2014, with persistent thrombotic occlusion of right pulmonary artery and severe pulmonary hypertension was referred for investigation of CTEPH. Patient was diagnosed as operable CTEPH with PVR 6,9 WU and underwent successful PEA. Histology of surgical material was compatible with the diagnosis of CTEPH.

Thirteen months after the PEA the patient deteriorated to FC III. RHC detected pulmonary valve gradient 35/21 mm Hg with area of 0.5 cm2 / m2. CT angiography showed penduculated tumor in the pulmonary artery. PET/CT showed no convincing signs of inflammation or neoplasia.

The patient underwent resection of pulmonary artery tumor in July 2016. Subsequent histology showed intimal sarcoma. Patient was indicated to palliative chemotherapy.

Conclusion: Pulmonary artery sarcoma should be considered as the differential diagnosis of CTEPH.

ICC17-2. EPIDEMIOLOGY AND OUTCOMES OF PATIENTS WITH CHRONIC THROMBO-EMBOLIC PULMONARY HYPERTENSION REFERRED FOR ENDARTERECTOMY BEFORE AND AFTER INITIATION OF ANGIOPLASTY IN THE FRENCH NATIONAL REFERENCE CENTER

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Balloon pulmonary arterial angioplasty (BPA) is an emerging technique for patients with chronic thrombo-embolic pulmonary hypertension (CTEPH) in whom surgical endarterectomy is not indicated or in non-operable patients.

Objective and Methods: This prospective registry study aimed to assess whether the implementation of the BPA program (February 2014) has modified the characteristics and outcomes of patients with CTEPH undergoing endarterectomy in the French National Reference Center. Indications of surgery or angioplasty were discussed by the multidisciplinary Heart Team. Comparisons were made using independent t-test or Chi-square test.

Results: Comparative characteristics of the 243 patients from the “pre-BPA” period (2012-2013) and the 246 patients from the “post-BPA” period (2015-2016) are presented in **Table.**

	Pre-BPA group N=243	Post-PBA group N=246	p value
Pre-operative characteristics			
Age (years)	61.4 ±15.0	60.0 ±14.2	0.28
Female sex	49.8%	47.2%	0.57
NYHA class III or IV	79.3%	73.5%	0.15
Mean PAP (mmHg)	44.6 ±12.0	45.9 ±11.7	0.23
Total Pulmonary Resistance (Wood Units)	13.7 ±5.0	10.6 ±5	0.37
Cardiac Index (L/min/m^2)	2.5 ±0.6	2.5 ±0.7	0.25
Post-operative characteristics			
Need of extra-corporeal support after surgery	6.6%	11.4%	0.06
Mechanical ventilation (days)	4.5 ±8.0	7.9 ±16.6	<0.01

Conclusions: The number and preoperative characteristics of patients with CTEPH undergoing endarterectomy remained comparable despite the emergence of angioplasty.

ICC17-3. LONGITUDINAL CARDIAC DEFORMATION IS REDUCED BOTH AT REST AND DURING EXERCISE IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AND REMAINS IMPAIRED AFTER PULMONARY ENDARTERECTOMY

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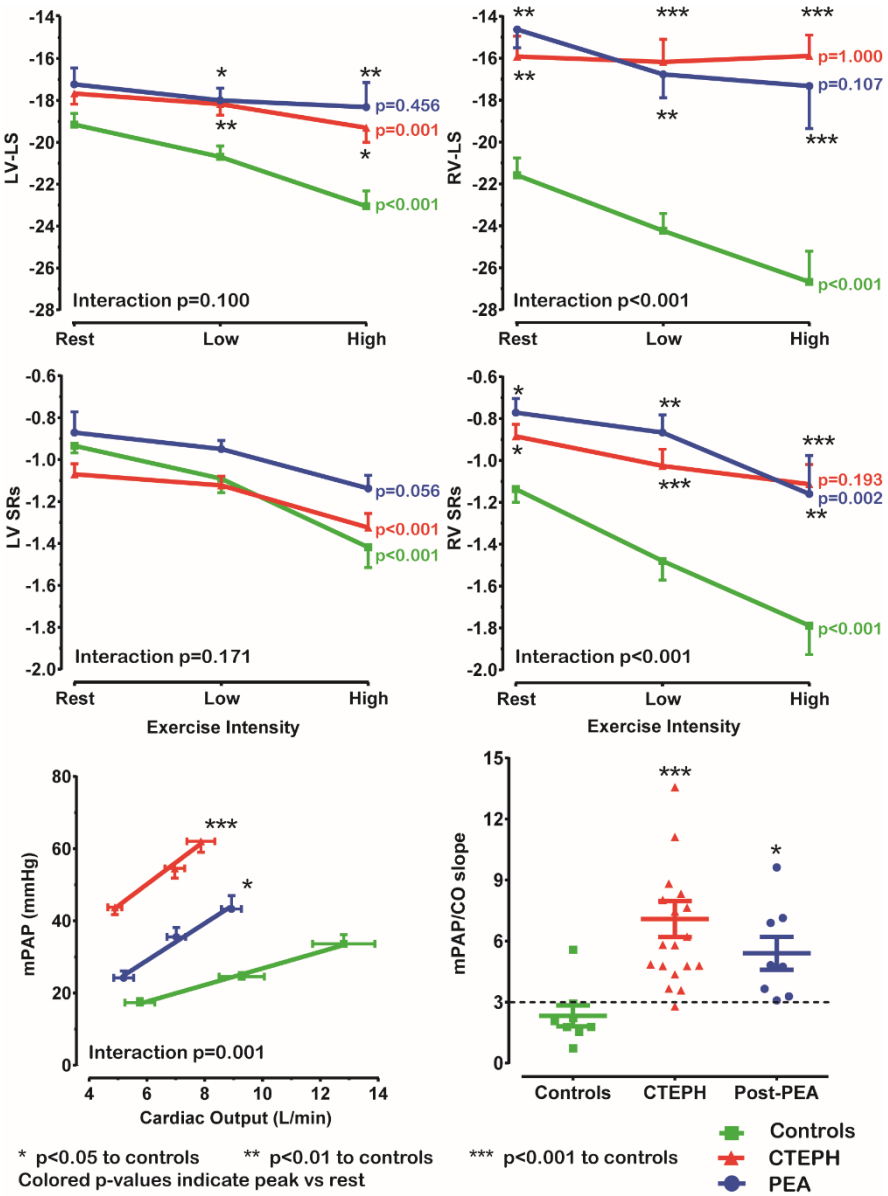
Background: Cardiac deformation imaging is predominantly performed at rest, yet patients are symptomatic with exercise and exercise capacity is an important prognostic factor in pulmonary hypertension.
Objective: To evaluate cardiac deformation at rest and during exercise in controls, patients with chronic thromboembolic pulmonary hypertension (CTEPH) and patients 6 months after pulmonary endarterectomy (PEA).
Methods: Nine controls, 22 CTEPH and 8 PEA patients underwent echocardiography at rest and during low and high intensity exercise on a semi-supine bicycle ergometer. Longitudinal strain (LS) and peak systolic strain rate (SRs) were derived from focused apical 4-chamber 2D grayscale images of the right (RV) and left (LV) ventricle. Cardiac output (CO) and mean pulmonary artery pressure (mPAP) were estimated by Doppler techniques.
Results: For LV LS and SRs the change with exercise was similar across the different groups (interaction p=0.1 and p=0.171). In contrast RV LS and SRs were significantly impaired at rest relative to controls (more positive) and the response to exercise was significantly different (interaction p<.001 for both). Notably RV LS and SRs augmented (more negative) in controls but while RV LS did not augment in either CTEPH or PEA patients, SRs augmented only modestly in PEA patients (see graphs). Exercise mPAP/CO relation differed between groups (p=0.001). An abnormal pulmonary vascular response (i.e. > 3mmHg/L/min) was observed in 94.7% (18/19) of CTEPH patients, 100% (8/8) of PEA patients and 12.5% (1/8) of controls (p<0.001).
Conclusion: RV, but not LV, LS and SRs are reduced in CTEPH patients and in patients after 'curative' PEA. These reductions are increasingly appreciable with exercise suggesting that exercise testing increases the sensitivity for detecting RV dysfunction.



ICC17-4. MULTICENTER, OBSERVATIONAL SCREENING SURVEY FOR THE DETECTION OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION FOLLOWING PULMONARY EMBOLISM

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Objectives:

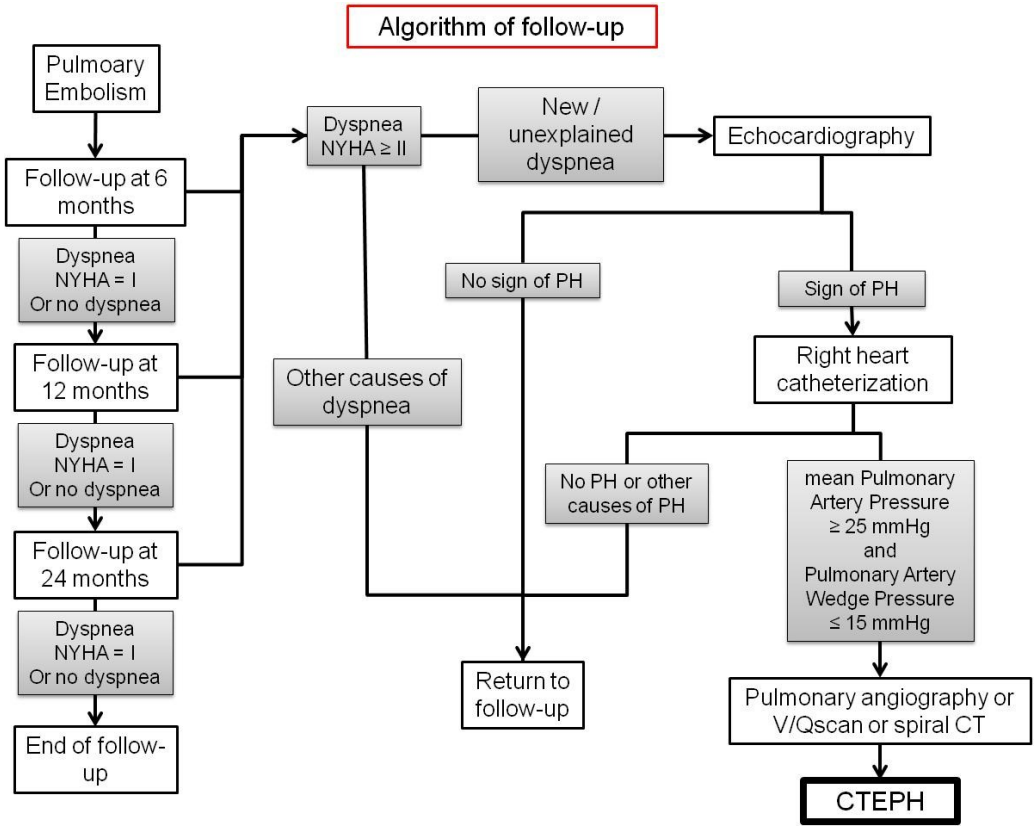
Primary objective

Evaluate the incidence rate of chronic thromboembolic pulmonary hypertension (CTEPH) following pulmonary embolism (PE)

Secondary objective

Test the usefulness of a dyspnea based screening algorithm for diagnosing CTEPH after PE

Methods: In a prospective, multicenter, observational phase V study, conducted between 2008 and 2015 we screened 1699 patients with PE in 11 Swiss pulmonary hypertension (PH) centers. 602 confirmed acute PE patients were included and followed over two years.



Results: Our main finding is a cumulative CTEPH incidence of 0.79% over two years. We identified 4 cases on 508 patients who completed the follow-up. The CTEPH incidence rate was 0.37% persons/year after PE in Switzerland. A match between the Swiss PH registry and our study database of all the 1699 screened patients strengthened our results. According to the registry, we missed no CTEPH case during our follow-up. Four additional cases occurred in the 1193 patients excluded or incompletely screened. Therefore, our protocol didn't segregate a higher risk population. The dyspnea based screening algorithm yielded a sensitivity of 100% and a specificity of 90.5%. Echocardiography in newly dyspneic patients showed a negative predictive value of 100%.

Conclusion: To our knowledge, this multicenter prospective study on CTEPH is one of the largest cohorts conducted in PE patients. The screening algorithm was able to detect all CTEPH cases occurring in the 2 years following PE. The low 0.79% incidence, when compared to the ~25 new diagnosed cases/year in Switzerland, raises the importance of silent embolic events in the CTEPH pathogenesis.

ICC17-6. LOW-GRADE SARCOMA ORIGINATING WITHIN THE THROMBOTIC MATERIAL IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH)

D. Di Pernaa, A. Sciortinoa, C. Giordanob, M. Concardib, M. Agozzinob, V. Graziolia, M. Pina, V.N. Merlia, B. Vaninia, A. Celentanoa, I. Parisia, R. Dorec, E. Arbustinib, A.M. D'Arminia

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Objectives: Patients with CTEPH complain exertion dyspnea, exercise intolerance, fatigue, occasional chest discomfort, syncope, hemoptysis and lower extremity edema. A similar phenotype is caused by pulmonary artery sarcomas of intimal and mural origin may show. The diagnosis of sarcoma can be incidental in PEA samples. We describe a unique case of low-grade intimal pulmonary sarcoma that originated within the thrombotic material of CTEPH.

Case description: A 45-yers old man developed deep vein thrombosis (DVT) of the right leg and of the left leg 5 years later; 2 years later he complained chest pain and dyspnea (WHO-II): a CT scan showed pleural thickening and effusion. The cytological examination of the pleural fluid excluded malignancy. Symptoms did not worsen but persist. One year later, following an accidental crash, an angio-CT scan of the neck and chest showed vertebral fractures (C6-7) and right CTEPH. The patient underwent uncomplicated right PEA.

The PEA samples showed a neoplastic proliferation in the context of the thrombotic layers. The immunophenotype was consistent with intimal sarcoma localized inside and entirely contained in thrombus. After PEA, the patient recovered; his WHO class declined from III to I; staging was negative. The patient participated in the decision of maintaining deep monitoring and not to perform pneumonectomy or radiation therapy or chemotherapy. One year later, imaging confirmed a good lung perfusion and excluded malignancy.

Conclusions: The one-year negative imaging follow-up supports our decision of monitoring the patient in a manner similar to that of the post-PEA in other patients with CTEPH.

ICC17-7. FAMILIAL CTEPH: A FAMILY WITH TWO AFFECTED MEMBERS TREATED WITH PULMONARY ENDARTERECTOMY (PEA)

D. Di Pernaa, V.N. Merlia, C. Giordanob, M. Concardib, T. Fiantaneseb, A. Sciortinoa, V. Graziolia, M. Pina, B. Vaninia, A. Celentanoa, I. Parisia, E. Arbustinib, A.M. D'Arminia

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Objectives: Recent case reports describe familial CTEPH in a sib pair[1] and in a mother and son[2]. The estimate of familial aggregation in CTEPH is complex because it should distinguish the family history of risk factors vs. those of CTEPH.

Methods: Since many years, we have implemented a genetic program to assess the familial risk of CTEPH. In this context, we identified a family in which two professional divers were diagnosed with CTEPH and underwent successful PEA at our institution.

Results: The proband is a 42-year-old man who had a first episode of dyspnea while practicing diving with the support of breathing apparatus. The patient was diagnosed with chronic bronchitis. A few months later, after a syncope, a thoracic-CT was given negative. However, the dyspnea (WHO IV) increased; echocardiography documented pulmonary hypertension (PAPs 105mmHg); an angioCT showed bilateral thrombo-embolism, later confirmed as CTEPH. The patient underwent bilateral PEA. He is now well, 4 years after surgery. One of his

paternal uncles, who shared with the proband the diving activity, also underwent successful PEA and is still doing well, 6 years after surgery. The pathology examination confirmed CTEPH in both samples series.
Conclusions: The occurrence of venous gas emboli and pulmonary edema are well-known complications in divers. Vice versa, the possible link between diving activity and PE is unexplored. Our case may appear anecdotal: however, we believe that all possible triggers and circumstances should be systematically included in the list of anamnestic factors to be explored in patients with CTEPH.

ICC17-8. INCIDENCE OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AFTER ACUTE PULMONARY EMBOLISM – A SINGLE CENTER EXPERIENCE

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Purpose: Although chronic thromboembolic pulmonary hypertension (CTEPH) is one of the most prevalent forms of pulmonary hypertension, it is still frequently underdiagnosed, and the true prevalence after acute pulmonary embolism (APE) is still undetermined. CTEPH is a potentially lethal condition and survivors of APE should be followed after the acute episode to detect signs or symptoms of CTEPH and determine the appropriate therapeutic strategy. The objective of this study was to analyze the association between CTEPH and APE with or without right ventricle dysfunction (RVD).
Methods: We studied consecutive 729 patients (398F, age 64±10.6yrs) with objectively proven APE: 208 pts with RVD (ICD10:I26.0,RVD(+) group) and 521 pts without RVD (ICD10:I26.9,RVD(-) group). Pts with known precapillary pulmonary hypertension were excluded. APE survivors were referred to our out-patient clinic for follow up, which also included CTEPH screening.
Results: All cause in-hospital mortality RVD(+) group reached 10%(21/208), while in RVD(-) it was 4,4%(23/521). From 685 survivors 314 pts completed at least 6 months follow-up (mean follow up 1.5±0.9yrs), while 361 others due to advanced age, severe comorbidities or distant residence were not controlled. Eventually, CTEPH was confirmed by right heart catheterization in 11(3,5%) of 314 pts. Importantly, 9(82%) CTEPH pts presented RVD during APE episode.
Conclusion: Since the incidence of CTEPH after episode of APE reached 3.5% in pts with at least 6 months follow-up and most of them presented RVD during APE, we suggest that clinical screening with subsequent imaging diagnosis should be performed in APE survivors especially with RVD at presentation.

ICC17-9. HIGH SENSITIVITY OF A NON-INVASIVE SCREENING STRATEGY FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AFTER ACUTE PULMONARY EMBOLISM

Y.M. Ende-Verhaar[1], D. Ruigrok[2], H.J. Bogaard[2], M.V. Huisman[1], L. Meijboom[3], A. Vonk Noordegraaf[2], F.A. Klok[1]
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Background: A non-invasive screening algorithm for chronic thromboembolic pulmonary hypertension (CTEPH) after acute pulmonary embolism (PE) was recently constructed. We aimed to evaluate the sensitivity of the algorithm and assessed the reproducibility of the individual items of the algorithm.
Methods: The algorithm was applied to 54 consecutive patients with confirmed CTEPH. Two independent researchers calculated the prediction score based on clinical characteristics at PE diagnosis, and evaluated the

ECG and NT-proBNP level assessed at CTEPH diagnostic work-up. Interobserver agreement for assessment of the prediction score, RV/LV ratio measurement on CTPA as well as ECG reading was evaluated by calculating kappa statistics.

Results: Median time between PE diagnosis and presentation with CTEPH was 8 months (interquartile range 5-13). 52 patients (96%, 95%CI 87-100%) had a high prediction score and/or CTEPH specific symptoms. The ECG/NT-proBNP combination was abnormal in 49 of 52 patients (94%, 84-99%). The sensitivity of the algorithm was 91% (79-97%), indicating that 27 of 30 cases of CTEPH would have been detected when applying the screening algorithm to 1000 random PE survivors with a 3% CTEPH incidence (projected negative predictive value 99.7%; 99.1-99.9%). The interobserver agreement for calculating the prediction score, RV/LV ratio measurement and ECG reading was excellent with a kappa of 0.96, 0.95 and 0.89 respectively.

Conclusion All components of the algorithm were highly reproducible. 91% of the CTEPH patients would have been identified by the algorithm, underlining its adequate sensitivity. Prospective validation of the algorithm in consecutive PE patients is required before clinical implementation.

ICC17-10. VENTILATION/PERFUSION SCINTIGRAPHY FOR THE DIAGNOSIS OF CTEPH AFTER SPLENECTOMY

M. Gerges, C. Gerges, G. Karanikas, W. Birkfellner, I. Schwarzinger, M. Publig, N. Skoro-Sajer,
R. Sadushi-Kolici, M. Frey, M. Schaffarich, J. Jakowitsch, M. Hacker, W. Klepetko, I.M. Lang

Department of Internal Medicine II, Division of Cardiology, Vienna General Hospital, Medical University of Vienna; Department of Biomedical Imaging and Image-guided Therapy, Division of Nuclear Medicine, Vienna General Hospital, Medical University of Vienna; Center for Medical Physics and Biomedical Engineering, Vienna General Hospital, Medical University of Vienna; Department of Laboratory Medicine, Division of Medical-Chemical Laboratory Diagnostics, Vienna General Hospital, Medical University of Vienna; Department of Surgery, Division of Thoracic Surgery, Vienna General Hospital, Medical University of Vienna

Background: Chronic thromboembolic pulmonary hypertension (CTEPH) results from non-resolving thrombi in the pulmonary vasculature. Patients after splenectomy are prone to complicated thrombosis and display a higher risk for venous thromboembolism and distal CTEPH. Radioisotopic ventilation-perfusion scanning (V/Q scan) is a safe and highly sensitive test detecting pulmonary thromboembolic disease. We sought to assess the diagnostic value of V/Q scan in splenectomized CTEPH patients.

Methods: In this prospective case control study we assessed V/Q scans of 77 patients with CTEPH, 10 of whom had undergone previous splenectomy. CTEPH was confirmed invasively in all patients by right heart catheterization and digital subtraction pulmonary angiography. V/Q scans were performed using 99mTc-labelled aerosol for ventilation and 99mTc-labelled 10-40µm human albumin microaggregates for perfusion. Images were evaluated for the presence of CTEPH using the revised PIOPED II criteria.

Results: The median time between splenectomy and V/Q scan was 14.9 years (range 3.8 – 34.3 years). V/Q scans in splenectomized CTEPH patients were normal in 4 cases (40%), showed a low probability in 3 cases (30%) and intermediate probability in 3 cases (30%). By contrast, V/Q scans in non-splenectomized CTEPH patients were high probability in 49 cases (73.1%). CTEPH patients with a history of splenectomy had significantly more normal and low probability V/Q scans (both $p<0.001$) than CTEPH patients without prior splenectomy.

Conclusions: V/Q scans may be negative or show a low probability for CTEPH in patients with a history of splenectomy.

ICC17-11. CAN WE SELECT THE PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION CANDIDATES FOR PULMONARY ENDARTERECTOMY ON THE BASIS OF MULTIDETECTOR COMPUTED TOMOGRAPHY ANGIOGRAPHY ONLY?

H. Gonzalez[1], M.J. Lopez Gude[2], MT. Velazquez Martin[1], Y. Revilla Ostolaza[3], S. Alonso Charterina[3], M. Perez Nunez[3], R. Morales Ruiz[3], J.L. Perez Vela[4], A. Albarran Gonzalez-Trevilla[1], C.A. Quezada Loaiza[5], C. Ortiz Bautista[1], N. Ochoa Parra[6], I. Ponz De Antonio[1], J.M. Cortina Romero[2], P. Escribano Subias[1]

[1-] University Hospital 12 de Octubre, Cardiology, Madrid, Spain; [2-] University Hospital 12 de Octubre, Cardiovascular Surgery, Madrid, Spain; [3-] University Hospital 12 de Octubre, Radiology, Madrid, Spain; [4-] University Hospital 12 de Octubre, Intensive Care Unit, Madrid, Spain; [5-] University Hospital 12 de Octubre, Pulmonology, Madrid, Spain; [6-] University Hospital 12 de Octubre, Investigation, Madrid, Spain

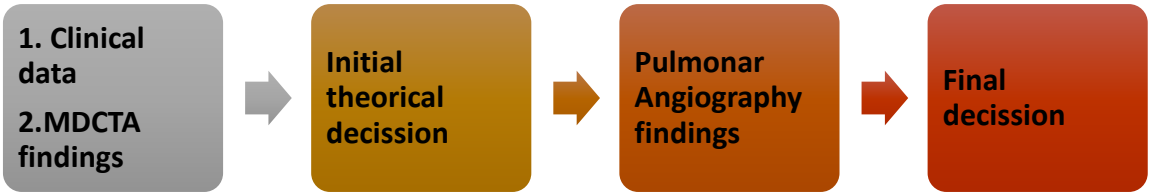
Background: Pulmonary Endarterectomy (PEA) is the treatment of choice. Operability is determined by accessibility of thrombi, comorbidities and clinical situation.

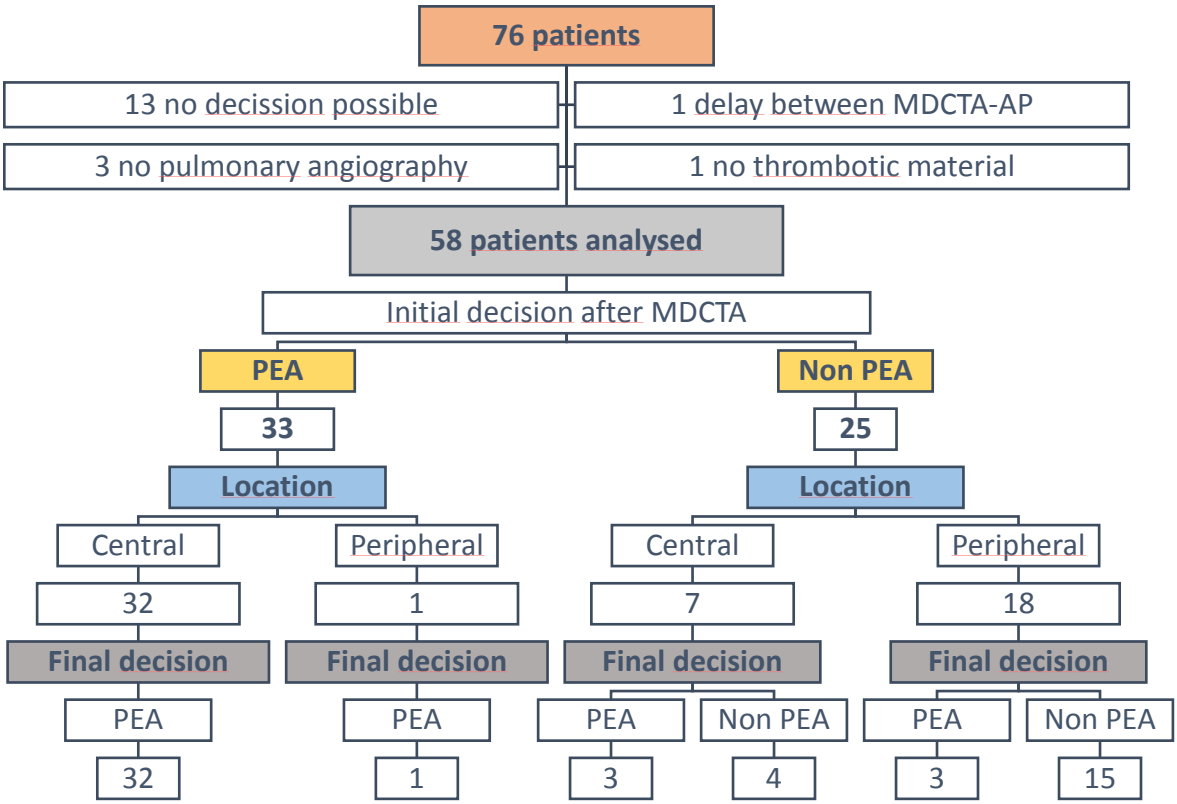
Purpose: evaluate current approach with Multidetector CT Angiography (MDCTA) and Pulmonary Angiography (PA) and propose a step-by-step approach based on MDCTA.

Methods: in 2016, we prospectively analyzed all patients evaluated in multidisciplinary session. They all underwent MDCTA and PA. Figure 1 shows session algorithm. Thrombi located from the pulmonary trunk to proximal segmental were central.

Results: 76 patients evaluated, 40 women (54,1%), age 57,9±1,7 years, 47 (62.7%) functional class III-IV, mPAP 48,3±1,6 mmHg. 18 patients excluded: 13 poor quality of MDCTA, 1 no thrombi, 3 no PA and in 1 important delay MDCTA-PA. The mean time between MDCTA and PA, 101±18 days. Of 58 analysed, 33 patients (57%) were candidates for surgery based on MDCTA (32 central and 1 peripheral). After evaluating the PA, the final decision was PEA in all patients. 25 were not suitable for PEA after analysing only MDCTA (7 central thrombi and 18 peripheral thrombi). After analyzing PA, from the group of central disease, they all were good candidates for PEA. From the group of peripheral thrombi, 3 patients were surgical. When studying the concordance between MDCTA and PA, we observed a good correlation (kappa index 0.75; $p<0.01$).

Conclusions: a step-by-step approach based on MDCTA can be applied. In our population, up to 57% PA could have been avoided. Nevertheless, those patients who have distal thrombi have to be evaluated with PA before rejected for PEA.





Correlation between MDCTA and PA			
PA	**MDCTA**	**Central**	**Peripheral**
Central		42	5
Peripheral		3	14
p < 0,001			

ICC17-12. UNUSUAL OBSTRUCTION OF THE PULMONARY ARTERIES

V. Grazioli, M. Pin, A. Sciortino, V.N. Merli, B. Vanini, A. Celentano, I. Parisi, G. Silvaggio, A.M. D'Armini

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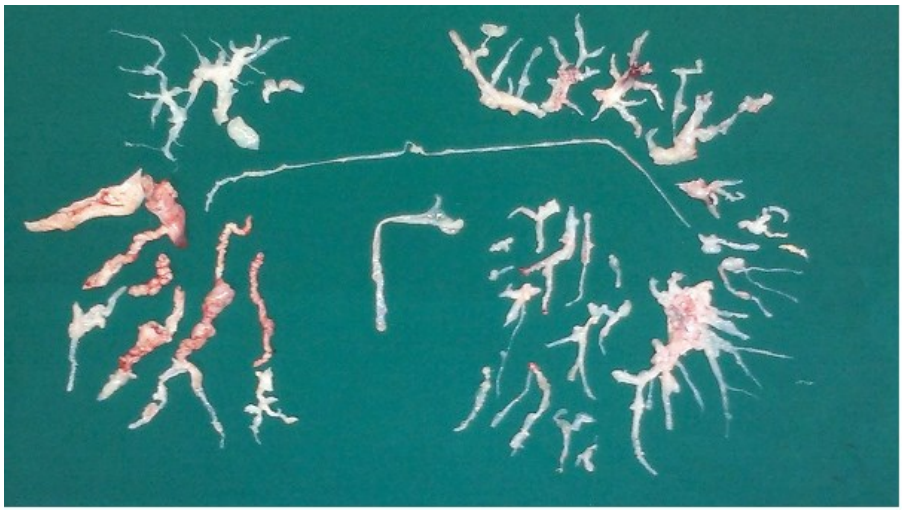
Objective: Pulmonary hypertension due to metastatic pulmonary arteries obstructions is an infrequent and fearsome disease with a very poor prognosis. It is generally misdiagnosed as a pulmonary thromboembolism. The life expectancy is related to the malignancy of tumor and to the development and progression of pulmonary hypertension.

Methods: We present an unusual case of 63-year old woman affected by an intravascular lung metastases of a mesenchymal bone knee tumor. After an accurately diagnostic work-up, the neoplastic origin of the lesions into the pulmonary arteries was suspected. The hemodynamical conditions registered with the right heart catheterization were severe: the median pulmonary arterial pressure (mPAP) was 49 mmHg and pulmonary vascular resistances (PVR) were 651 dyne*sec*cm^{-5}. A bilateral pulmonary endarterectomy was performed with multiple circulatory arrests and reperfusion periods, moderate hypothermia (24°C) and ventricular fibrillation.

Results: An improvement of the clinical status of the patient was obtained after surgery: the reduction of mPAP and PVR registered were respectively 59% and 71%. The amelioration of the clinical condition after surgery

allowed to treat the patient also with an adjuvant chemotherapy to reduced the tumor progression. The patient died two years after surgery for metastatic cerebral lesions.

Conclusions: Considering the malignancy of the primary neoplasm and the metastatic presentation, a curative and radical treatment is usually difficult to obtain. Thank to this multidisciplinary approach, an improvement of the survival was attained.



Reconstruction of the tumor material removed from the pulmonary arteries.
The material removed from the pulmonary arterial branches, from the right atrium and inferior vena cava.

ICC17-13. ROLE OF S100A4 IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Objective: We recently reported on the role of the Receptor for Advanced Glycation Endproducts(RAGE) in patients with chronic thromboembolic pulmonary hypertension(CTEPH). S100A4, an intra- and extracellular calcium-binding protein, interacts with RAGE, but is also a marker for the epithelial mesenchymal transition, as is the epithelial growth factor receptor (EGFR). We sought to investigate the role of S100A4 and EGFR in patients with CTEPH. Results were compared to patients with idiopathic pulmonary arterial hypertension (iPAH) and aortic valve stenosis (AVS).

Methods: We recruited 37 CTEPH patients undergoing pulmonary endarterectomy (PEA), 19 iPAH patients undergoing lung transplantation(LuTX) and 15 patients with AVS undergoing aortic valve replacement(AVR). Thirty-nine healthy volunteers served as controls. Immunoassays were performed on serum samples collected before and after surgery.

Results: S100A4 serum concentrations were significantly decreased in iPAH (mean±standard error mean: 1.3±0.2ng/ml; p=0.002) and CTEPH (1.4±0.1ng/ml; p<0.001) compared to controls (2.1±0.1ng/ml). There was no significant difference in patients with AVS (2.3±0.2ng/ml) when compared to controls (p=0.435). EGFR serum concentrations of iPAH (38.6±2.2ng/ml), CTEPH (38.5±2.5ng/ml) and AVS (37.4±3.2ng/ml) patients were

significantly decreased in comparison to healthy volunteers (47.4±1.7ng/ml; p=0.006, p=0.004, p=0.006, respectively).

Conclusions: Further investigations into the pathophysiologic role of S100A4 and EGFR in patients with CTEPH and iPAH are warranted.

ICC17-14. RISK FACTORS FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH) – IMPORTANCE OF THYROID DISEASE

L. Hobohm[1,2], V. Krieg [1], C. Liebetrau[3], S. Guth [4], S. Koelmel[1], K. Pohl [1], C. Troidl [5], L. Essmann[5], H. Rossmann[6], S. Konstantinides[1], E. Mayer[4], C. Wiedenroth[4]*, M. Lankeit[1,7]*

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Objective: To investigate the prevalence of risk factors with a special focus on thyroid disease in patients with chronic thromboembolic pulmonary hypertension (CTEPH).

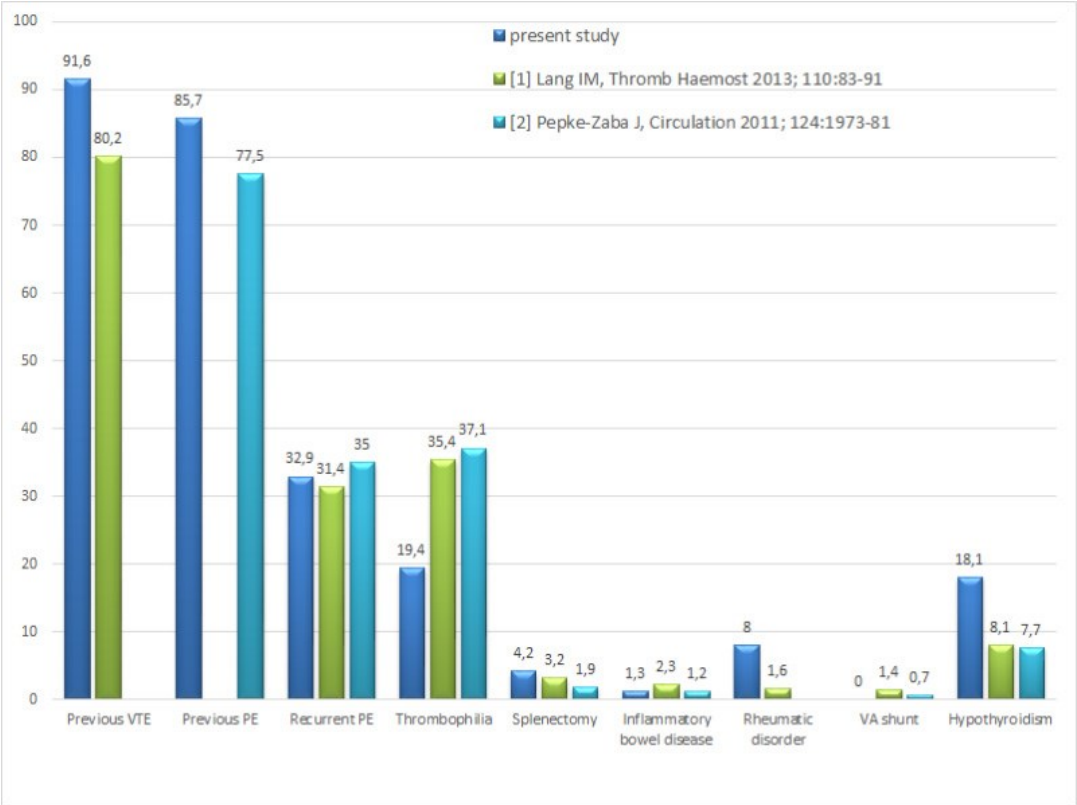
Methods: Patients with operable CTEPH treated with pulmonary endarterectomy (PEA) at a single referral centre between 01/2014 and 12/2015 were studied. In a subgroup of 122 patients (51.5%), TSH, fT3, fT4 and TPO antibodies were measured in venous blood samples taken before PEA.

Results: Overall, 237 patients (median age, 62 [52-72] years; 46.0% female; 78.5% NYHA class III/IV; mean PA pressure, 43 [34-50] mmHg) were studied. Almost all patients reported a history of venous thromboembolism (91.6%); of those, the majority had a pulmonary embolism (PE; 85.7%) and 32.9% more than one PE event. Risk factors are shown and compared to previous cohort studies in **Figure 1**.

Fifty-five patients (23.2%) had a known thyroid disease (18.1% hypothyroidism; 5.1% hyperthyroidism) and of those, 90.9% received specific treatment. fT4 levels correlated weakly with cardiac output (r=-0.191; p=0.039) and were higher in patients who died in-hospital (1.4[1.2-1.5] vs 1.1[1.0-1.2]ng/dl; p=0.037) or during follow-up (1.4[1.1-1.6] vs 1.1[1.0-1.6]ng/ml; p=0.016). However, thyroid disease was not associated with outcomes.

Conclusions: In 237 operable CTEPH patients, a history of VTE and thyroid disease were more common than previously reported. fT4 levels were higher in patients who died in-hospital or during long-term follow-up.

Figure 1. Frequency of risk factors for CTEPH



ICC17-15. PROANGIOGENIC AND WOUND HEALING MOLECULAR AND HISTOLOGICAL FINGERPRINT OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

D. Iloska[1], J. Wilhelm[2], S. Guth[3], R. Savai[1,2], R. Schermuly[2], L. Fink[2,4], W. Seeger[1,2], E. Mayer[3], S.S. Pullamsetti[1,2]

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Objective: Chronic thromboembolic pulmonary hypertension (CTEPH) as a multifactorial disease, ultimate to death, if unrecognized and untreated. The aim of the project is to identify the transcriptional regulatory landscape in CTEPH and compare the histopathological and molecular imprint with idiopathic PH (IPAH).

Methods: Laser capture microdissection of vessels was performed on *ex-vivo* CTEPH, IPAH and donor lung tissues followed by microarray screening. In addition, from these lungs, vascular remodeling and collagen content were quantified. Total vessel density was evaluated by vWf and CD62P immunoreactivity. Comparable analysis was implemented on proximal and distal pulmonary endarterectomy (PEA) material.

Results: Morphometric analysis confirmed similar extent of medial hypertrophy, yet differences in collagen deposition and vascularization between CTEPH and IPAH lungs, as compared to donors. Bioinformatics analysis of the microarray data revealed differently (607) and commonly (366) regulated genes and gene networks. Our *in vitro* studies, verified that two of the differently regulated genes in CTEPH i.e. CHI3L1 and ENPP2 (Chitinase 3-Like-1 and Ectonucleotide pyrophosphatase/phosphodiesterase 2, respectively) promote neovascularization and migration of vascular cells. In the distal PEA tissues, elevated levels of angiogenic markers and an increased

insoluble collagen were noted as compared to proximal PEA tissues. Importantly, microarray analysis suggests deregulation of several important transcription factor networks in the distal PEA tissue, suggesting their contribution to neovascularization and collagen synthesis.

Conclusion: These studies highlight the activation of neoangiogenic processes in CTEPH, rather than in IPAH. Further studies will provide insights into the molecular mechanisms and can possibly contribute to identify novel therapeutic targets.

ICC17-16. QUALITY OF LIFE AMONG CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION PATIENTS IN RUSSIAN FEDERATION

A. Klimenko, N. Shostak, V. Shemenkova

Pirogov Russian National Research Medical University, Department of Faculty Therapy named after A.I. Nesterov, Russian Federation, Moscow

Objective: to assess the physical and mental state of patients with chronic thromboembolic pulmonary hypertension (CTEPH) using the «SF-36 Health Status Survey».

Materials and methods: 15 patients with a confirmed diagnosis of CTEPH: 7 men (46,7%) and 8 women (53,3%). The average age of the patients was 60,1 ± 3,7 years. Patients completed the SF-36 quality-of-life questionnaire, assessed the extent of pulmonary hypertension, a six-minute walk test (6MWT) to determine the functional class (FC), the duration of symptoms and other.

Results: the quality of life (QoL) indicators for women were lower in all scales than in men, but the reliability of differences was revealed only on the scale of physical functioning (p = 0,033), reflecting the degree of restriction of exercise. Analysis of the components of QoL, depending on the clinical data showed that the higher FC and the shorter the distance in 6MWT, the lower the level of physical and mental functioning, the level of systolic pressure in the pulmonary artery correlates only with the index of physical functioning. The duration of the disease also affects the value of QoL indicators, patients during the 1-st year of treatment rated their general health condition much higher than patients with longer periods of treatment (p = 0,026).

Discussion: SF-36 quality of life questionnaire is an actual, simple, accessible tool for assessing the main components of physical and mental health in patients with CTEPH in real clinical practice.

ICC17-17. MIXED VENOUS OXYGEN SATURATION CAN BE A PROGNOSTIC FACTOR OF HEMODYNAMIC STABILITY IN FOLLOW-UP PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Objective: Follow-up catheterization study is usually performed on the patients with chronic thromboembolic pulmonary hypertension (CTEPH) who were received balloon pulmonary angioplasty (BPA). We sometimes experience the hemodynamic deterioration in follow-up phase. While, a prognostic factor of hemodynamic stability is unclear. The purpose of this study is to clarify the prognostic factor of hemodynamic stability after BPA in patients with CTEPH.

Method: We retrospectively analyzed 22 patients with CTEPH (age 62.8±13.6 year-old, 15 female). All patients were received BPA. Their mean pulmonary arterial pressure (mPAP) were improved below 30mmHg (26.9±3.0mmHg). In follow-up catheterization study, hemodynamic stability was maintained in 15 patients, while hemodynamic deterioration was found in 7 patients. Hemodynamic deterioration was defined as elevation of mPAP in follow-up study and/or

requirement of additional BPA. We retrospectively analyzed their hemodynamic and respiratory data from immediately before follow-up.

Result: There were statistically significant differences in age and mixed venous oxygen saturation (SvO_2) between stable and worsening patients (58.8±13.8 vs 71.5±8.2, 70.6±4.7 % vs 64.3±6.0%; $p<0.05$, respectively). In multivariate analysis, independent prognostic factor of hemodynamic stability was SvO_2 (OR,1.308; 95%CI, 1.001 to 1.710; $p<0.05$). Receiver operating characteristic curve demonstrated SvO_2 level as predictor of hemodynamic stability in follow-up phase with area under the curve of 0.81. An SvO_2 cut-off value above 68.5% had a 73% sensitivity and an 86% specificity.

Conclusion: In follow-up patients with CTEPH, SvO_2 can be a prognostic factor of hemodynamic stability after BPA.

ICC17-18. SCREENING OF AUTO-ANTIBODIES IN PATIENT WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH) WITH PROTOARRAY® ANALYSIS

A. Naito[1], N. Tanabe[1,2], T. Hiwasa[3], T. Jujo[1,2], T. Sugiura[1], S. Sakao[1], K. Tatsumi[1]

[1-] Department of Respirology, Graduate School of Medicine, Chiba University; [2-] Department of Advanced Medicine in Pulmonary Hypertension, Graduate School of Medicine, Chiba University; [3-] Department of Biochemistry and Genetics, Graduate School of Medicine, Chiba University

Objective: It has been reported that some circulating auto-antibodies have been detected in patients with cardiovascular diseases. However, the auto-antibodies in CTEPH patients were not well investigated so far. Here we investigated the circulating auto-antibodies in CTEPH patients using ProtoArray® and alpha-LISA analysis.

Method: *Subjects*

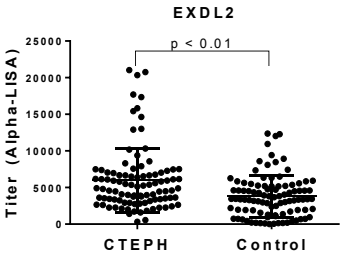
We collected the serum samples from CTEPH patients who were diagnosed with CTEPH at our institute from 2001 to 2015. Healthy donors (HDs) were collected from those who underwent annual medical checkup.

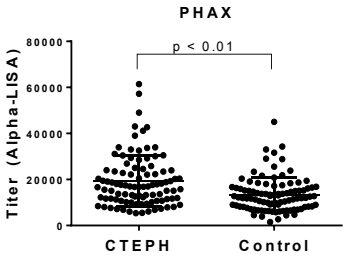
Measurement of the auto-antibody titer

As a screening study, serum samples from 5 CTEPH patients and 5 HDs were profiled on ProtoArray® Human Protein Microarrays containing more than 9,000 human proteins. Pick-upped auto-antibodies were evaluated in serums from 96 CTEPH patients and 96 HDs using Amplified luminescence proximity homogenous assay (Alpha LISA).

Result: We pick-upped 63 auto-antibodies of which CTEPH patients have high titers from the result of ProtoArray® analysis. Measuring the titers of those auto-antibodies using Alpha LISA analysis, we found that CTEPH patients have high titers of Exonuclease 3'-5' Domain Containing 2 (EXDL-2) antibody (CTEPH group;5997±4371, HD group; 3801±2842) and Phosphorylated Adaptor For RNA Export (PHAX) antibody (CTEPH group;19261 ± 11182, HD group; 13371±7386). Those titers were not correlated with the hemodynamic parameters measured by right heart catheter.

Conclusions: CTEPH patient had high titers for the two antibodies. Although the detail function of those antibodies remained unclear, further investigation might give us the information about the etiology of CTEPH.





ICC17-19. THE EFFECT OF MULTIDISCIPLINARY TEAM APPROACH ON THE IMPLEMENTATION RATE OF MAJOR DIAGNOSTIC AND THERAPEUTIC PROCEDURES OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH)

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Objective: We performed the study to identify the effect of multidisciplinary team (MDT) approach on the implementation rate of major diagnostic and therapeutic procedures that are essential for managing chronic thromboembolic pulmonary hypertension (CTEPH).

Methods: We retrospectively analyzed the medical records of patients with CTEPH from December 1994 to December 2016. We divided the period into pre- and post-MDT era based on November 2013, the date of establishment of MDT in our hospital. We compared the implementation rate of major diagnostic and therapeutic procedures between the eras.

Results: Among 109 patients with CTEPH, mean age was 53.3 and 58 (53.2%) were male. Thirty-four (31.2%) patients were diagnosed at post-MDT era. For the diagnostic procedures, the implementation rates of pulmonary angiography and right heart catheterization were significantly increased at post-MDT era (20.0% [n=15] vs. 97.1% [n=33], p<0.001 and 12.0% [n=9] vs. 97.1% [n=33], p<0.001, respectively). For the therapeutic procedures, the implementation rates of pulmonary endarterectomy and balloon pulmonary angioplasty were significantly increased after the establishment of MDT (33.3% [n=25] vs. 64.7% [n=22], p=0.002 and 0.0% [n=0] vs. 26.5% [n=9], p<0.001, respectively). The rate of patients treated with medication only was significantly decreased at post-MDT era (66.7% [n=50] vs. 11.8% [n=4], p<0.001).

Conclusion: The MDT is associated with increased implementation rates of major diagnostic and therapeutic procedures of CTEPH, especially those are essential but hard to be performed due to technical barriers including pulmonary angiography, right heart catheterization, pulmonary endarterectomy and balloon pulmonary angioplasty.

Table 1. Implementation rates of major diagnostic procedures

	Pre-MDT era	Post-MDT era	p-value
Patients with CTEPH, No. (%)	75 (68.8)	34 (31.2)	
V/Q scan, No. (%)	70 (93.3)	32 (94.1)	> 0.999
Pulmonary angiography, No. (%)	15 (20.0)	33 (97.1)	< 0.001
CT angiography, No. (%)	65 (86.7)	34 (100.0)	0.029
Echocardiography, No. (%)	74 (98.7)	34 (100.0)	> 0.999
Right heart catheterization, No. (%)	9 (12.0)	33 (97.1)	< 0.001

Abbreviations: MDT = Multidisciplinary team; CTEPH = Chronic thromboembolic pulmonary hypertension; V/Q = Ventilation/Perfusion; CT = Computed Tomography.

Table 2. Implementation rates of major therapeutic procedures

	Pre-MDT era	Post-MDT era	p-value
Patients with CTEPH, No. (%)	75 (68.8)	34 (31.2)	
PEA, No. (%)	25 (33.3)	22 (64.7)	0.002
BPA, No. (%)	0 (0.0)	9 (26.5)	< 0.001
Medical treatment only, No. (%)	50 (66.7)	4 (11.8)	< 0.001

Abbreviations: MDT = Multidisciplinary team; CTEPH = Chronic thromboembolic pulmonary hypertension; PEA = Pulmonary endarterectomy; BPA = Balloon pulmonary angioplasty.

ICC17-20. THE FIRST CASE OF AN EXTRASKELETAL MYXOID CHONDROSARKOMA IN THE PULMONARY ARTERY

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Objective: We present the first case of a primary extraskeletal myxoid chondrosarkoma (EMC) in the pulmonary artery (PA).

Methods: A 21-year old female presented herself with progressive dyspnea. The diagnostic work up with CT scan showed a mass in the main trunk of the PA. Complete work up included echocardiography, positron-emission tomography, computed tomography, a heart MRI and V/Q scan of the lung and an immunological screen. Cytology of EBUS FNA showed myxoid proliferations, suggestive of a mesenchymal neoplasia

Results: A pulmonary tumor endarterectomy with partial reconstruction of the right pulmonary artery in deep hypothermia without circulatory arrest was performed. The postoperative course was uneventful and she was discharged at day 6.

The definitive histological examination revealed an EMC, usually found in the lower proximal extremities. A translocation of the EWSR1-gen in the q22;q12 locus was found as well as S100 negativity. Microscopic complete resection was proven in the distality of the endarterectomy specimen.

No adjuvant therapy was performed. 14 months after surgery the patient is free of recurrence in follow-up CT scan and back to her normal work life and sports activity.

Conclusions: EMC are a rarity within the mesenchymal tumors with an incidence of 2-3 % of all soft-tissue sarcoma and most likely occur in the proximal lower extremities. Prognosis is good with 5 year survival rate of about 90 % after complete resection. Localization in the PA has not been described so far.

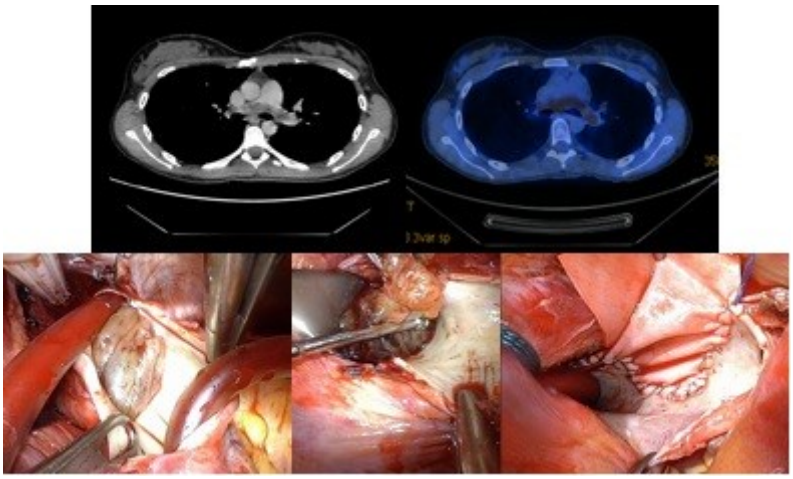

Figure: Contrast enhanced CT scan of the chest (A), PET/CT scan shows a limited FDG uptake (B), Tumor is located in the main PA (C) extending into the left and right trunk (D), where it is attached to the wall and was completely resected and right PA partially reconstructed (E).

ICC17-21. VALUE OF DUAL-ENERGY CT DERIVED IODINE MAPS FOR THE ASSESSMENT OF REGIONAL LUNG PERFUSION IN CTEPH COMPARED TO V/Q SCAN USING CT PULMONARY ANGIOGRAPHY AS A STANDARD OF REFERENCE

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Objective: To assess the value of dual-energy CT pulmonary angiography (DECT) in patients with chronic thromboembolic pulmonary hypertension (CTEPH) compared to V/Q Scan.

Methods: From 09/2013 to 07/2016 12 CTEPH patients (6 females, median age 71 years) underwent DECT and V/Q scan. Iodine maps derived from DECT and axial and coronal reformations were interpreted. V/Q scan and DECT were assessed segment-wise by two radiologists independently using a 4 point Likert scale (0=normal; 3=no perfusion). Vascular involvement assessed on CT pulmonary angiography (CTPA) served as standard of reference, using a 4 point Likert scale (0=normal vessels; 3=occlusion of major segmental vessels). Interobserver agreement between readers (Cohen's kappa) and diagnostic accuracy were calculated. Correlations of scores were compared using Spearman's rho. P<.05 was considered statistically significant.

Results: Interobserver agreement was higher for DECT compared to scintigraphy (kDECT=.65 versus kScinti=.45). Analysis of ROC curves showed that AUC for detection of perfusion deficiency were .727 and .711, respectively for DECT with p<.001 for both and .58 and .58 for scintigraphy with p=.037 and .044, respectively. DECT showed higher sensitivity and specificity compared to V/Q scans for detection of low perfused segments (sensitivity: 74% vs. 69%; specificity: 69% vs. 44%). Ratings of DECT and CTPA showed moderate (r=.46; p<.001) and ratings of scintigraphy and CTPA showed weak correlation (r=.23; p=.038).

Conclusions: In our study, assessment of lung perfusion with DECT shows higher accuracy and is more robust compared to V/Q scans in patients with CTEPH, and might replace V/Q scans in the future.

ICC17-22. SINGLE CENTRE PROSPECTIVE STUDY OF PATIENTS WITH CHRONIC THROMBOEMBOLIC DISEASE (CTED)

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Introduction: Diagnostic criteria and treatment are established in patients with CTEPH. It is less clear what to do in the context of CTED without pulmonary hypertension at rest. This is increasingly recognized as a cause of symptoms and exercise limitation post-pulmonary embolism.
Methods: Between January 2015-2017 176 locally referred Papworth patients with suspected CTED/CTEPH underwent a standard assessment as per international guidelines. CTED was defined as symptomatic patients with technically operable disease and mPAP<25 mmHg. Patients diagnosed with CTED had an incremental cardiopulmonary exercise test (CPET) and exercise RHC at 40% maximum workload. Symptoms were measured by CAMPHOR questionnaire. All patients were assessed at the pulmonary endarterectomy (PEA) MDT.
Results: Of 176 patients, 35(20%) were diagnosed with CTED. Airway obstruction was present in 14(40%). Peak oxygen consumption and symptoms correlated with VE/VCO_2 at anaerobic threshold (p= 0.007, p=0.003 respectively). Cardiac output increase on exercise at the time of RHC correlated with exercise capacity (p=0.019). 17% of CTED patients underwent PEA. There were no significant changes in exercise capacity, symptoms, echocardiographic parameters or NT pro-BNP at one-year follow-up in the non-operated group.
Conclusions: CTED accounts for a significant proportion of referrals to our center. The majority of CTED patients were not operated on but remained clinically stable during a one-year period. Coexisting cardiorespiratory disease needs careful assessment. The only haemodynamic measurement associated with symptoms and exercise capacity was increase in cardiac output on exertion.

ICC17-24. HYPOCAPNIA AS A PROGNOSTIC MARKER OF CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Rationale: Excessive ventilation and desaturation during exercise are hallmarks of chronic thromboembolic pulmonary hypertension (CTEPH). The increased dead space and inequalities in ventilation/perfusion distribution, associated to abnormal stimuli that reduced $PaCO_2$ to sub-physiological levels can contribute to hyperventilation. Hypocapnia is established marker of prognosis in pulmonary arterial hypertension (PAH). **Objective**: To evaluate the prognostic value of hypocapnia in CTEPH patients.
Methods: Fifty patients with nonoperable CTEPH, residual pulmonary hypertension after pulmonary endarterectomy or not operated by refusal (49±15 yrs, 66% female) were consecutively evaluated from September 2010 to August 2016. At baseline, patients performed arterial blood gas analysis, right heart catheterization and six-minute walk test.
Results: During a median follow-up of 4.2 years (95% CI, 2.8-5.6), 15 deaths were recorded. Estimated survival was 85% at 1 year (95% CI, 80-91); 80% at 2 yrs (95% CI, 74-86) and 77% at 3 yrs (95%CI, 71-83). At univariate analysis, mortality was associated with NYHA functional class III and IV [HR 10.2 (1.3-78), p 0.025], cardiac index (CI) ≤ 1.8L/min/m^2 [HR 3.4 (1.2-9.4), p 0.017], pulmonary vascular resistance (PVR) ≥ 1000dynes.s.cm^{-3} [HR 6.3 (1.7-22.6), p 0.005], pulmonary artery compliance ≤ 0.6mL/mmHg [HR 7.4 (2.0-26.4), p 0.002], 6 minute walk distance ≤ 360m [HR 4.2 (1.1-15.3), p 0.03] and

PaCO2 ≤ 31mmHg [HR 5.1 (1.4-18.1), p 0.012]. At bivariate analysis including CI and $PaCO_2$, PaCO2 ≤ 31mmHg was an independent risk factor for mortality [adjusted HR 3.9(1.08-14.8), p 0.039].

Conclusion: Hypocapnia at baseline is a marker of mortality in not operated CTEPH patients.

ICC17-25. INSPIRATORY MUSCLE WEAKNESS CONTRIBUTES TO EXERTIONAL DYSPNEA IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Rationale: Patients with CTEPH often present with symptoms of exertional breathelessness and decrease in exercise tolerance. Determination of potentially-reversible factors contributing to dyspnea remains an unmet clinical need in CTEPH.

Objective: To evaluate the influence of inspiratory muscle weakness (IMW) on exercise capacity and dyspnea during effort in patients with CTEPH.

Methods: Thirty-nine patients with CTEPH (48 ± 15 yrs, 61% female), confirmed by right heart catheterization, underwent an incremental cardiopulmonary exercise test, 6-minute walk test and maximum inspiratory pressure (MIP) measurement.

Results: MIP < $70\%_{pred}$ was found in 46% of patients. On a multiple linear regression analysis, including pulmonary vascular resistance (PVR) and New York Heart Association functional class (NYHA FC), MIP was independently associated with 6MWD and $\dot{V}O_{2\ PEAK}$. In comparison to patients with preserved inspiratory muscle strength, those with IMW presented stronger sensations of dyspnea throughout exercise, even when adjusted for ventilation ($p<0.05$).

Conclusion: IMW is associated with dyspnea and poor exercise capacity in patients with CTEPH.

Medical treatment and interventional treatment

ICC17-5. TRANSITIONING FROM SILDENAFIL TO RIOCIGUAT IN PATIENTS WITH PERSISTENT OR INOPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION IMPROVES EXERCISE CAPACITY AND HEMODYNAMICS

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Objectives: The purpose of the study was to assess hemodynamic and functional parameters in patients with inoperable or persistent chronic thromboembolic pulmonary hypertrnsion [CTEPH] after the medication change from sildenafil to riociguat when the drug became commercialy available.

Methods: We enrolled 24 patients with the diagnose of inoperable or persistent CTEPH confirmed by CTEPH-team. All subjects previously treated at least 3 months with sildenafil dosed 25mg or higher three times daily were switched to riociguat therapy in gradually escalated doses. The assesstment of WHO functional class, 6-minute walk distance [6MWD], and invasive hemodynamics were performed in all subjects in three time points: before the beginning of sildenafil therapy, during the sildenafil therapy and 3 to 6 months after the final range of riociguat was reached. No balloon pulmonary angioplasty was performed during the period of drugs replacement and follow up.

Results: After the medication change from sildenafil to riociguat significant increase of cardiac output (4,2±0,9 vs 4,6±0,8 l/min; p=0,03), significant decrease of pulmonary vascular resistance (9,7±4,8 vs 8,5±3,9 Wood u.; p=0,03), signicant improvement of 6MWD (274±177 vs 316±131 m; p=0,04) were observed. Moreover, significant improvement in WHO functional class was also noticed (p<0,05).

Conclusions: Replacement sildenafil by riociguat in patients with inoperable or persistent CTEPH significantly reduces pulmonary vascular ressistance, increases cardiac output, improves 6MWD and WHO functional class.

Non-operable or persistent CTEPH (n=24)							
Hemodynamics	NO (baseline)	NO vs SIL p	SIL	SIL vs RIO p	RIO	NO vs RIO p	**SIL vs RIO mean % of change**
HR [u/min.]	78,7 ± 26,0	0,94	78,3 ± 16,6	0,16	81,3 ± 20,1	0,43	-
SAP [mmHg]	129,2 ± 18,0	0,77	128,6 ± 17,7	< 0,01	117,4 ± 12,8	< 0,01	- 9%
DAP [mmHg]	78,9 ± 11,3	0,11	75,7 ± 11,1	< 0,01	67,4 ± 8,6	< 0,01	- 11%
mRAP [mmHg]	8,8 ± 4,3	0,47	8,2 ± 3,9	0,87	8,2 ± 3,9	0,50	-
sPAP [mmHg]	89,3 ± 18,0	0,22	84,1 ± 18,7	0,18	80,5 ± 22,4	0,07	-
dPAP [mmHg]	34,4 ± 9,9	0,03	30,9 ±10,7	0,04	29,0 ±10,3	< 0,01	- 6%
mPAP [mmHg]	55,3 ± 12,3	0,04	49,7 ± 13,1	0,19	47,9 ± 14,0	0,01	-
PCWP [mmHg]	10,7 ± 3,1	0,44	11,2 ± 2,6	0,02	10,1 ± 2,76	0,45	- 10%
CO [l/min]	4,2 ± 1,0	0,64	4,2 ± 0,9	0,03	4,6 ± 0,8	0,04	+ 9%
CI [l/min*m^2]	2,4 ± 0,6	0,85	2,4 ± 0,6	0,04	2,6 ± 0,5	0,04	+ 8%
PVR [Wood U.]	11,1 ± 3,6	0,13	9,7 ± 4,8	0,03	8,5 ± 3,9	< 0,01	- 13%
SVR [Wood U.]	20,6 ± 4,7	0,22	19,4 ± 5,0	<0,01	16,4 ± 3,4	< 0,01	-16%

Functional assesment and NT-proBN							
6MWT [m]	-	-	274 ± 177	0,04	316 ± 131	-	+ 14%
NT-proBNP [pg/ml]	-	-	1774 ± 2068	0,18	1362 ± 1248	-	-
Functional capacity (WHO)							
I	0		1		2		
II	2		6		7		
III	18	<0,05	17	<0,05	15	<0,01	-
IV	4		0		0		

NO – *without any specific treatment*, SIL – *treated by sildenafil*, RIO – *treated by riociguat,* HR – *heart rate*, SAP – *systolic arterial pressure*, DAP – *diastolic arterial pressure*, mRAP – *mean right atrial pressure*, sPAP – *systolic pulmonary artery pressure*, dPAP – *diastolic pulmonary artery pressure*, mPAP – *mean pulmonary artery pressure*, PCWP – *pulmonary capillary wedge pressure*, CO – *cardiac output*, CI – *cardiac index*, PVR – *pulmonary vascular resistance*, SVR – *systemic vascular resistance*, 6MWT – *six minutes walk test*, NT-proBNP – *N-terminal pro b-type natriuretic peptide*

ICC17-23. EFFICACY OF PULMONARY HYPERTENSION-TARGETED DRUGS FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AFTER PERCUTANEOUS TRANSLUMINAL PULMONARY ANGIOPLASTY

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Objectives: The purpose of this study was to investigate the effectiveness of PH-targeted drugs for CTEPH after PTPA.

Methods: This study retrospectively included 128 patients with CTEPH who underwent PTPA by August 2016. Fifty-nine patients discontinued PH-targeted drugs on the basis of attaining less than 25mmHg of mean pulmonary artery pressure (PAP) after PTPA. Hemodynamic parameters were compared among baseline, after PTPA and after withdrawal of PH-targeted drugs.

Results: The median age, dilated vessels per person and observational period from the final PTPA to the withdrawal of drugs was 64[58-72] years old, 16[12-20] vessels and 362[268-496] days, respectively.
The number of patients with each PH-targeted drug was broken down to 51(86%) in phosphodiestrase-V inhibitors, 50(80%) in endothelin receptor antagonists and 3(5%) in riociguat. The number of patients with combination medical therapy was 49(80%). There was no significant difference in PAP between after PTPA and after withdrawal of drugs. However, cardiac Index(CI) and Pulmonary vascular resistance(PVR) were significantly worsened after withdrawal of drugs (baseline vs. after PTPA vs. after withdrawal of drugs; PAP: 38[32-43] vs. 19[16-22] mmHg vs. 20[16-23], P<0.01; PVR: 6.4[4.8-10.4] vs. 1.9[1.5–2.7] vs. 3.0[2.1-3.7], P<0.01, CI: 2.6[2.2–3.1] vs. 3.0[2.6–3.4] vs. 2.7[2.4-3.0], P<0.01).

Conclusions: It may be possible to safely discontinue PH-targeted drugs after an improvement of the hemodynamics to normal range with PTPA, but small degree of small vessel disease persists after PTPA, aggravating pulmonary hemodynamics after withdrawal of PH-targeted drugs, which thus affecting cardiac function in addition to pulmonary vasculature.

ICC17-26. RESIDUAL RIGHT VENTRICULAR DYSFUNCTION AND REMODELING AFTER BALLOON PULMONARY ANGIOPLASTY IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: PREVALENCE, CLINICAL CHARACTERISTICS AND PREDICTORS

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Objective: Balloon pulmonary angioplasty (BPA) improves hemodynamics, right ventricular (RV) function and prognosis in patients with chronic thromboembolic pulmonary hypertension (CTEPH). Residual RV remodeling is one of the remaining issues, which may influence clinical outcome and treatment strategy. This study aimed to clarify the frequency and characteristics of residual RV remodeling after BPA.

Methods: We retrospectively included 68 consecutive CTEPH patients who underwent BPA along with cardiovascular magnetic resonance (CMR). Residual RV remodeling was defined as RV end-diastolic volume (EDVI)>100ml/m^2 and RV ejection fraction (EF)<45%. We divided all patients into residual RV remodeling (RRR) and normalized RV remodeling (NRR) group using CMR at 3-month follow-up after BPA series.

Results: BPA significantly improved clinical parameters on average. We categorized patients into RRR (56%) and NRR group (44%). At baseline, RRR group had higher proportion of male, higher BNP level, more prolonged QRS duration. RRR group showed significantly larger RVEDVI and worse RVEF at both baseline and follow-up. At follow-up, WHO functional status in RRR group was significantly worse than that in NRR group, although no death and hospitalization was observed. No significant hemodynamics differences were observed in both groups at follow-up. On multivariable logistic regression analysis, male, prolonged QRS and lower RVEF at baseline were independently associated with residual RV remodeling after BPA.

Conclusions: We found high incidence of unrecognized residual RV remodeling with worse symptoms at short-term follow-up after BPA albeit hemodynamics improvement. Male, prolonged QRS and lower RVEF at baseline were risk factors of residual RV remodeling after BPA.

ICC17-27. BALLOON PULMONARY ANGIOPLASTY FOR CTEPH: 3-YEARS EXPERIENCE FOCUSED ON SAFETY AND EFFICACY IN 128 PATIENTS

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Objectives: to report safety and efficacy of balloon pulmonary angioplasty (BPA) in CTEPH since the beginning of our experience in february 2014.

Methods: we rewieved :

1 - procedural complications in 128 patients (706 sessions) : complications was classified in 3 types : « mechanical » (distal perforation, dissection, rupture), reperfusion pulmonary injury (RPI)using Inami classification and « others » (renal insufficiency, local hematoma, …).

2 – clinical and hemodynamic follow-up at 6 monthes : NYHA class, 6 minutes walking distance, hemodynamic datas (mean PAP, cardiac index, pulmonary vascular resistance).

Results: they are summarized in the following tables :

Complications	% sessions	% patients
Mechanical	6,2%	30%
RPI	7%	32,5%
Others	1%	5,1%
Deaths related to BPA	0,5%	3%

Mean PAP is the strongest predictor of RPI in multivariate analysis. Mechanical complications and RPI were more frequently observed in the first half of patients.

FOLLOW-UP	Baseline	Follow-up	p value
NYHA	2,81 ± 0,6	1,68 ± 0,6	< 0 ,001
6MWT	397 ± 137	446 ± 125	< 0 ,001
Mean RAP	8,2 ± 3,8	7 ± 4,5	0,063
Mean PAP	44,7 ± 10, 4	29,7 ± 9	< 0 ,001
Cardiac index	2,62 ± 0,6	3,01 ± 0,6	< 0 ,001
PVR	8,1 ± 3,1	3,6 ± 1,5	< 0 ,001

Conclusions: BPA is effective in improving clinical conditions and pulmonary hemodynamic in patients presenting inoperable CTEPH, but serious complications are not so infrequent. This invasive must be performed in experienced centers having all the means necessary to manage this patients.

ICC17-28. WARFARIN PHARMACOGENETICS IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH)

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Objective: The genotypes of cytochrome-P450 (CYP) 2C9 variants (**2,*R144C and **3,*I359L) and vitamin K epoxide reductase complex 1 (VKORC1 c.−1639G>A) influence warfarin dosing and are incorporated in algorithms used to predict stable warfarin dose. They explain 10% to 45% of the overall variance. We investigated the genotypes of P450 (CYP) 2C9 and VKORC1 in a consecutive series of patients with CTEPH who underwent PEA.

Methods: The clinical series is constituted of 365 patients, 177 males and 188 females, median age 55 (IQR:41-66). All patients had CTEPH and were treated with oral anticoagulation at individually adjusted doses. All patients signed an informed consent for genetic testing. We investigated the Minor Allele Frequency (MAF) of *VKORC1 (c.-1639G>A), CYP2C9*2 (R144C;rs1799853)* and *CYP2C9*3 (I359L;rs1057910)*. The MAF was compared with that of EU population as reported in large public databases [Exome Variant Server-EVS; 1000Genomes(1K); EXAC].

Results: The genotypes are reported in the table. MAF was similar in our CTEPH cohort and in EU population.

	GENOTYPE(366 PATIENTS)			MAF(732 alleles)		
	GG	GA	AA	MAF in CTEPH	MAF EU POPULATION (1K/EXAC/EVS)	p-value
VKORC1 (c.-1639G>A)	130	169	67	0.41	0.38/-/-	p=0.7
CYP2C9*2 (R144C)	269	90	7	0.14	0.12/0.09/0.13	p=0.57
CYP2C9*3 (I359L)	302	62	2	0.09	0.07/0.06/0.066	P=0.47

The levels of INR were within ranges (2-3.5) in 231/366 patients (63%). Genotypes and MAF were not significantly different in patients with INR in range vs. out-of-range.

Conclusions: At present, pharmacogenetics does not explain the individual differences in INR (range or out of range) in chronically anti-coagulated patients with CTEPH.

ICC17-29. PREOPERATIVE MONOTHERAPY WITH PHOSPHODIESTERASE-5 INHIBITOR, ENDOTHELIN RECEPTOR ANTAGONIST, OR RIOCIGUAT DOES NOT AFFECT OUTCOMES AFTER PULMONARY THROMBOENDARTERECTOMY

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Objective: Use of pulmonary arterial hypertension (PAH)-targeted oral medical therapy prior to pulmonary thromboendarterectomy (PTE) is common among patients referred to our center. Whether the use of various classes of these medications affects outcomes from this procedure has not been adequately investigated.
Methods: In order to assess class effects on PTE outcomes, we reviewed consecutive patients referred to UCSD for PTE and compared patients on no PAH-targeted medical therapy to those treated with monotherapy with either phosphodiesterase type 5-inhibitors (PDE5i), endothelin receptor antagonists (ERA), or soluble guanylate cyclase stimulator (sGCS). Patients on combination therapy were excluded. A multivariate logistic regression was completed to examine the effects of monotherapy with one of the three classes on outcomes from PTE surgery (including mortality, residual pulmonary hypertension with PVT >400 dyn.sec.cm^{-5}, and reperfusion lung injury).
Results: Of the 521 patients operated on between 2013 and 2015, 48.0% were on pre-operative PAH-targeted medical therapy. After excluding those on combination therapy, there were 80 patients on PDE5i, 68 patients on sGCS, and 16 on ERA. In a multivariate logistic regression model which adjusted for age, sex, BMI, right atrial pressure, PVR, cardiac index, history of VTE, and disease level, there was no significant increase in the odds a patient pre-treated with any of the three classes of PAH-targeted oral medications on post-operative mortality, residual pulmonary residual pulmonary hypertension, reperfusion lung injury or pulmonary hemorrhage (Table 1).
Conclusions: Preoperative monotherapy with PDE5i, sGCS, or ERA does not significantly increase the incidence of any important morbidity or mortality after PTE surgery.

Table 1. Multivariate Logistic Regression for Single Treatment Effect on Various Post PTE Outcomes

	Number of Patients	Mortality OR (95% CI)	Residual PH OR (95% CI)	Reperfusion Lung Injury OR (95% CI)	Pulmonary Hemorrhage OR (95% CI)
No Drug	271	Reference	Reference	Reference	Reference
PDE5i	80	1.552 (0.202-8.187)	2.332 (0.862-6.221)	1.869 (0.954-3.601)	1.773 (0.651-4.550)
sGCs	68	NA	1.845 (0.619-5.147)	1.598 (0.763-3.223)	1.029 (0.277-3.089)
ERA	16	5.414 (0.233-52.172)	2.377 (0.320-11.593)	1.845 (0.464-6.108)	NA

PTE = pulmonary thomboendarterectomy; PH = pulmonary hypertension; RPE = reperfusion edema; PDE5i = phosphodiesterase type 5 inhibitor; sGCs = soluble guanylate cyclase stimulator; ERA = endothelin receptor antagonist (Of note, there were no deaths among patients on sGCs alone and no episodes of pulmonary hemorrhage among patietns on ERA alone so OR cannot be calculated for either of these groups.)

ICC17-30. RIOCIGUAT FOR INOPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: RESULTS FROM A PHASE II LONG-TERM EXTENSION STUDY

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Objectives: In a 12-week Phase II trial of patients with pulmonary arterial hypertension (n=33) and inoperable chronic thromboembolic pulmonary hypertension (CTEPH; n=42), riociguat was well tolerated and improved 6-minute walking distance (6MWD) and World Health Organization functional class (WHO FC). We assessed the tolerability and efficacy of riociguat in the subgroup of patients with inoperable CTEPH in the long-term extension (LTE) phase of the study.
Methods: During this multicenter, open-label, uncontrolled LTE study (NCT00454558), riociguat dose was adjusted at the physician's discretion (range of 0.5–2.5 mg tid). The primary outcome was safety and tolerability; secondary outcomes included 6MWD, WHO FC, and survival.
Results: Forty-one patients with inoperable CTEPH entered the LTE, with 23 (56%) remaining on riociguat at the final cut (median treatment duration 77 months). The most common adverse events were nasopharyngitis (59%) and peripheral edema (39%). No patients experienced hemoptysis. At Month 48, mean±SD 6MWD had improved by 59±82 m (n=27), and WHO FC had improved/stabilized/worsened versus baseline in 46/46/7% of patients (n=28). Four-year survival was 91% and 26 (63%) patients experienced a clinical worsening event. The most common event was starting a new PH treatment (46%).
Conclusions: Long-term use of riociguat was safe and well tolerated in patients with inoperable CTEPH. Improvements in 6MWD and WHO FC were maintained at 4 years in those patients continuing treatment. These data support riociguat as a long-term treatment option for inoperable CTEPH.

ICC17-31. FOCUSED BALLOON PULMONARY ANGIOPLASTY FOR TRULY NON-OPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION – THE UK EXPERIENCE

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Objective: Balloon Pulmonary Angioplasty (BPA) is emerging as a viable treatment for Chronic Thromboembolic Pulmonary Hypertension (CTEPH), although the potential benefits and safety of BPA have not been fully described in a truly non-operable cohort of patients, carefully selected at a surgical multi-disciplinary team meeting. We assessed the clinical results of a focused BPA approach, treating disease largely confined to basal sub-segmental arteries (A8-10), in patients with distal, non-operable CTEPH.
Methods: Sixteen (7%) of the total referred patients with CTEPH were selected for BPA from October 2015 to February 2017. Preoperative assessment included right heart catheterisation, cardiopulmonary exercise test (CPET), distance covered on a 6-minute walk test (6MWT), pro-NT brain natriuretic peptide (pro-NT BNP) level and RV function and dimensions by transthoracic echocardiography. These measurements were repeated 3-months (where possible) after completion of treatment and compared.
Results: Forty-five procedures were performed in 16 patients (mean age 64, 11 male, median 3 procedures per patient) treating 89 vessels (Left A8 – n=16, A9 – n=14, A10 – n=13, other – n=2; Right A8 – n=16, A9 – n=8, A10 –

n=14, other – n=6). We observed 4 minor complications (3 femoral haematoma and 1 radiological-evidence of lung reperfusion injury not requiring oxygen therapy) but no major complications or deaths. Baseline and 3-month data (Table 1) indicate improvements in haemodynamic, exercise capacity and RV function indices.
Conclusions: Focused BPA, targeting lesions in the basal sub-segmental pulmonary arteries that receive the most pulmonary blood flow, achieves haemodynamic improvement and clinically important benefits with relatively few treatments required per-patient and few complications.

Variable	Baseline	+ 3-months*	p-value
RAP, mmHg	8.8±4.0	5.6±2.3	**<0.001**
RVEDP, mmHg	11.5±6.1	7.1±2.5	**0.02**
mPAP, mmHg	44.0±11.7	34.1±7.9	**0.004**
CO_T, L/min	4.6±1.1	4.9±1.3	0.18
PVR_T, Dynes.cm^{-5}	623±239	411±142	**0.001**
VO_2 max, ml/min/kg	17.5±5.0	17.8±3.8	0.45
6MWT, metres	399±113	455±97	**0.02**
Pro-NT BNP, pg/mL	1087±1162	441±689	0.07
TAPSE, cm	1.9±0.5	2.2±0.5	0.44
RV mid-diameter, cm	4.4±1.1	3.2±0.2	0.45

RAP – right atrial pressure, RVEDP – right ventricular end diastolic pressure, mPAP – mean pulmonary artery pressure, CO_T – cardiac output by thermodilution, PVR_T – pulmonary vascular resistance by thermodilution, VO_2 max – maximum oxygen consumption, 6MWT – 6-minute walk test; TAPSE – Tricuspid annular plane systolic excursion.

Table 1: Pulmonary haemodynamics, exercise capacity and echocardiographic / biomarker assessment of RV function at baseline and 3-months after BPA (* not all 16 patients have completed 3-month follow-up at the time of submission).

ICC17-32. EFFICACY OF PTPA FOR LONG-TERM OUTCOME IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Purpose: The purpose of this study was to investigate the 3-year outcome and efficacy in pulmonary hemodynamics after PTPA.
Methods: This study retrospectively included 180 patients with inoperable CTEPH patients who underwent PTPA until October 2016. Firstly, we investigated the 3-year survival and restenosis. Secondary, hemodynamic parameters such as mean pulmonary arterial pressure (PAP), pulmonary vascular resistance (PVR) at baseline, after short-term (0.5-1.5 years), mid-term (1.5-3.5 years) and long-term (> 3.5 years)
Results: A median of age, PTPA session performed per person and dilated vessels per person was 66[55-74] years old, 4[2-4] and 15[10-20] respectively. 1-year, 3-year, and 5-year survival (all-cause) were 98.9% (95% confidence interval [CI], 94.9-99.7), 98.1% (95% CI, 93.7-99.3), and 95.8% (95%CI, 85.9-98.6), respectively. Time-course changes of hemodynamics after percutaneous transluminal pulmonary angioplasty (PTPA) in patients with chronic thromboembolic pulmonary hypertension. Mean pulmonary arterial pressure (PAP) was least squares mean of 37.9 mmHg (95%CI, 36.6-39.3) at baseline, and changed to 22.3 mmHg (95%CI, 20.8-23.7) at short-term, 20.8 mmHg (95%CI, 19.1-22.5) at mid-term, and 19.3 mmHg (95%CI, 16.5-22.0) at long-term after PTPA (all P<0.001 vs.

baseline, respectively). Pulmonary vascular resistance (PVR) was 7.9 Wood units (95%CI, 7.4-8.4) at baseline, and changed to 2.9 Wood units (95%CI, 2.4-3.5) at short-term, 2.9 Wood units (95%CI, 2.3-3.6) at mid-term, and 2.4 Wood units (95%CI, 1.3-3.5) at long-term after PTPA (all P<0.001 vs. baseline, respectively).
Conclusions: The findings of the present study demonstrate that long-term favorable outcomes after PTPA can be expected in patients with CTEPH.

ICC17-33. EFFICACY AND SAFETY OF MACITENTAN FOR INOPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: RESULTS FROM THE RANDOMIZED CONTROLLED MERIT STUDY

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Objective: The randomized, double-blind MERIT study (NCT02021292) evaluated the endothelin receptor antagonist macitentan in primary inoperable CTEPH.
Methods: Patients in WHO functional class (FC) II–IV with a pulmonary vascular resistance (PVR) ≥400 dyn·sec/cm^5 and 6-minute walk distance (6MWD) of 150-450 m were randomized 1:1 to placebo or macitentan 10 mg once daily and treated for 24 weeks. Inoperability/eligibility was assessed by an independent adjudication committee. Phosphodiesterase type-5 inhibitors (PDE-5i) or oral/inhaled prostanoids were allowed for patients in WHO FC III–IV. The primary endpoint was PVR at Week 16, expressed as percent of baseline. The main secondary endpoint was mean change from baseline to Week 24 in 6MWD. Exploratory endpoints included mean change from baseline to Week 16 in cardiac index (CI) and mean right atrial pressure (mRAP), and percent of baseline N-terminal pro B-type natriuretic peptide (NT-proBNP) at Week 24.
Results: 80 patients were randomized to macitentan (n=40) or placebo (n=40). At baseline, 61% were receiving PAH therapy; 96% PDE-5i. Compared with placebo, PVR and 6MWD significantly improved in the macitentan-treated group (Table). The treatment effect on PVR and 6MWD was similar regardless of baseline PAH treatment. Exploratory endpoints also improved in the macitentan-treated group (Table). Most common adverse events (macitentan versus placebo) were peripheral edema (22.5% vs. 10.0%) and decreased hemoglobin/anemia (17.5% vs. 2.5%). Five patients (placebo) prematurely discontinued treatment. Two patients (placebo) died.
Conclusion: In the MERIT study, macitentan led to significant improvements in cardiopulmonary hemodynamics and exercise capacity in inoperable CTEPH patients and was well tolerated.

Table

	Baseline (N=40) Mean (SD)		Post-baseline* (N=40) Mean (SD)		% of baseline Geometric mean (95% CL)		Treatment effect (Ratio)** (95% CL) p value
	Placebo	Macitentan	Placebo	Macitentan	Placebo	Macitentan	
PVR, dyn·sec/cm^5 (primary endpoint)	984 (487)	929 (380)	899 (476)	723 (454)	87.2 (78.5, 96.7)	73.0 (63.6, 83.8)	0.84 (0.70, 0.99) p=0.04
NT-proBNP, pg/mL	1793 (2075)	2204 (2943)	1433 (1450)	1553 (2252)	90.9 (76.5, 107.9)	72.6 (61.2, 86.2)	0.79 (0.63, 0.99) p=0.04
	Baseline (N=40) Mean (SD)		**Post-baseline* (N=40) Mean (SD)**		**Change from baseline Mean (SD)**		**Treatment effect (Difference)** (95% CL) p value**
	Placebo	**Macitentan**	**Placebo**	**Macitentan**	**Placebo**	**Macitentan**	
6MWD, m (secondary endpoint)	351 (74)	353 (88)	352 (121)	388 (83)	1.0 (83.2)	35.0 (52.5)	34.0 (2.9, 65.2) p=0.03
mRAP, mmHg	8.7 (5.2)	10.4 (8.1)	8.6 (5.3)	7.8 (5.9)	−0.1 (3.5)	−2.6 (8.4)	−1.47 (−3.77, 0.84) p=0.21
CI, L/min/m^2	2.2 (0.6)	2.2 (0.5)	2.2 (0.5)	2.6 (0.8)	0.0 (0.4)	0.4 (0.7)	0.43 (0.18, 0.67) p<0.001

*Post-baseline is Week 16 for PVR, mRAP and CI, and Week 24 for 6MWD and NT-proBNP
**From ANCOVA model adjusted by variable at baseline as covariate

ICC17-34. COMPLICATIONS WITH THE USE OF THE NEW ORAL ANTICOAGULANT DRUGS IN THE SETTING FO CTEPH TREATMENT – REPORT OF TWO CASES

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Introduction: We describe two cases of our cohort with complications regarding anticoagulation management and intra-operative difficulties where the common factor was the use of one of the new oral anticoagulant drugs (NOAC).

Case 1: Female, 38 year old, with *situs inversus*, diagnosed with CTEPH, and she was in use of rivaroxaban on the last 5 months.

submitted to pulmonary endarterectomy, being surgery succesfull and post surgery recovery as expected.

She was discharged with Warfarin, as our routine for post surgery anticoagulation, with INR of 3, and with good exercise recovery.

After 1 month, as she lives in a remote place and had difficulties in INR controll, the cardiologist with our agreement changed her to rivaroxaban once more.

After 4 months of surgery she returns to ER with dyspnea, and Angio CT showed a segmental accute pulmonary embolism (PE).
Six months after surgery another acute PE, confirmed with Angio CT to be almost all lower lobe.

Case 2: Male, 48 year old, with no co-morbidities and confirmed diagnosis of CTEPH, referred to us for surgery evaluation. He was already in use of rivaroxaban because of his pulmonologist preference, and reporting mild worsening od exercise capacity in the last 6 months.
admitted at the emergency department with acute worsening of symptoms, and a new echocardiogram showed presence of thrombotic material at the main pulmonary artery, causing major obstruction.
at surgery lot of acute and sub-acute clots were found, making surgery more difficult because of the volume they make inside the vessel, making the dissection harder.

ICC17-35. SAFTEY AND EFFICACY OF OUR CURRENT STRATEGY OF BALLOON PULMONARY ANGIOPLASTY FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Objective: In performing balloon pulmonary angioplasty (BPA) for CTEPH patients, we originally tried to fully dilate two lesions in initial procedure and repeated procedures for other lesions. Since 2014, we totally changed our BPA strategy. Currently, we dilate as many lesions as possible with smallest balloon in initial procedure and then optimize the dilation of the lesions with bigger balloon in following procedures. We investigated difference of outcome between previous and current strategies.
Methods: One hundred forty three CTEPH patients were enrolled. Patients divided into 2 groups according to the date of their initial procedure (previous group; 83 patients who underwent initial BPA before November 2014, current group; 60 patients who underwent initial BPA after November 2014). Efficacy and safety of BPA in 2 groups were investigated. Efficacy outcome were improvement of mean pulmonary artery pressure (mPAP) and cardiac index (CI) after fourth BPA procedures. Safety outcome was frequency of severe complications after BPA needing intratracheal intubation or extracorporeal membrane oxygenation (ECMO).
Results: Significant deference was not observed in efficacy outcome between previous and current groups (mPAP: -16.9 ± 1.6 vs. -17.6 ± 1.6 mmHg, p=0.37, CI: +0.42 ± 0.14 vs. +0.19 ± 0.12 L/min/m^2, p=0.10). Intratracheal Intubation and ECMO were needed in 8 patients (9.6%) and 6 patients (7.2%) in previous group, whereas none of the patients in current group experienced severe complications.
Conclusions: Our current strategy could improve the safety of BPA while keeping the efficacy of BPA.

ICC17-36. BALLOON PULMONARY ANGIOPLASTY FOR PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: A JAPANESE MULTICENTER REGISTRY

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Objective: Balloon pulmonary angioplasty (BPA) is recently reported to be effective for selected patients with chronic thromboembolic pulmonary hypertension (CTEPH). However, all reports are results from a relatively small cohort treated at a single center. We conducted a retrospective multicenter registry to evaluate safety and efficacy of BPA.

Methods: A total of 308 patients who underwent BPA between November 2004 and March 2013 were registered at 7 institutions in Japan. Data were retrospectively analyzed to evaluate clinical outcome and complications. Survival from the initial BPA session was evaluated.

Results: Hemodynamic parameters were significantly improved in 249 patients in whom all sessions were completed. Mean pulmonary arterial pressure decreased by 19 mmHg after final BPA. In 196 patients who underwent right heart catheterization at follow-up, improvement of hemodynamic parameters was maintained. Complications occurred in 36.6% of a total of 1408 sessions. Most of them were related to procedure: reperfusion pulmonary injury (17.8%), hemoptysis (14.0%), and pulmonary artery perforation (2.9%). Death occurred in 12 patients during follow-up. The leading causes of death were right heart failure, multi-organ failure, and sepsis. Overall survival was 99.6% at 1 year and 98.4% at 3 years after the initial BPA session.

Conclusions: The first multicenter registry in Japan revealed that BPA performed at experienced pulmonary hypertension centers is a safe and effective therapeutic option in patients with CTEPH. Further investigation is needed to confirm long-term safety and efficacy of BPA.

ICC17-37. RIOCIGUAT FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH): 2-YEAR RESULTS FROM THE CHEST-2 LONG-TERM EXTENSION

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Objective: Riociguat significantly improved 6-minute walking distance (6MWD) and other efficacy endpoints in patients with inoperable or persistent/recurrent CTEPH in the 16-week CHEST-1 trial. We present 2-year data from the CHEST-2 long-term extension.

Methods: The primary endpoints were safety and tolerability; secondary endpoints included 6MWD, WHO functional class (FC), *N*-terminal prohormone of brain natriuretic peptide (NT-proBNP), survival, and clinical worsening-free survival. Kaplan–Meier analyses and a Cox proportional-hazards regression model were used to assess correlation between efficacy parameters and long-term outcomes.

Results: Of 243 patients who completed CHEST-1, 237 (98%) entered CHEST-2. At this cut-off (March 2014) most patients had been treated for ≥2 years. Riociguat was well tolerated and no new safety signals were identified. Serious adverse events occurred in 129 (54%) patients (8% considered drug-related) and 14 (6%) patients discontinued therapy because of adverse events. Mean±SD 6MWD improved from CHEST-1 baseline by 50±68 m

(n=162) and WHO FC improved/stabilized/worsened in 39/58/3% of patients (n=170). 2-year survival and clinical worsening-free survival rates were 93% and 82%, respectively. 6MWD and NT-proBNP at CHEST-1 baseline and Week 16 correlated significantly with long-term survival and clinical worsening-free survival, and WHO FC correlated with clinical worsening-free survival.

Conclusion: Riociguat has a good long-term safety profile and is the first pharmacologic therapy for CTEPH to show sustained clinical effect. The correlation of 6MWD and NT-proBNP with long-term survival emphasizes the prognostic value of these endpoints in patients with CTEPH.

ICC17-38. POOR SUBPLEURAL PERFUSION AS A PREDICTOR OF FAILURE AFTER BALLOON PULMONARY ANGIOPLASTY FOR NON-OPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Background: It has been shown that 'poor subpleural perfusion' (PSP) in the capillary phase of pulmonary angiography (PA), which is related to microvasculopathy in chronic thromboembolic pulmonary hypertension (CTEPH), predicts poor outcome after pulmonary endarterectomy (Tanabe, Chest 2012). We assessed the association between PSP and failure of Balloon Pulmonary Angioplasty (BPA) in 101 non-operable CTEPH.

Method: Subpleural perfusion in the capillary phase of PA was classified as normal or poorly perfused as previously described. Patients were divided according to hemodynamic results after the last BPA: a failure group (defined as mean PAP >30mmHg and PVR decrease <20% after BPA [n=11]) or a success group [n=90].

Results: Baseline characteristics and hemodynamics were similar in the two groups (table). In contrast, PSP was observed in 45.5% of patients in the failure group versus 15.6% in the success group (p=0.016). Multivariate analysis revealed that PSP was the only predictor of BPA failure (Odds Ratio 4.524, 95% confidence interval 1.213-16.877, p=0.025).

Conclusion: Poor subpleural perfusion in the capillary phase of pulmonary angiography, suggesting the presence of diffuse microvasculopathy, is associated with significant residual pulmonary hypertension after BPA. In our experience, it affected approximately 10% of non-operable CTEPH who underwent BPA.

Variable	Success group n=90	Failure group n=11	p value
Baseline			
Age (years)	63.5 ± 13.8	62.7 ± 14.5	0.867
Male (n, %)	47 (52.2)	7 (63.6)	0.474
NYHA Fc (I,II / III,IV) (%)	26.5 / 73.5	27.3 / 72.7	0.369
6 minutes walk distance (m)	401 ± 118	380 ± 92	0.627
mean PAP (mmHg)	45.6 ± 10.2	45.6 ± 11.4	0.988
Cardiac Index (L/min/m^2)	2.51 ± 0.48	2.66 ± 0.80	0.393
PVR (wood unit)	8.39 ± 3.25	7.58 ± 2.28	0.422
Poor subpleural perfusion	14 (15.6%)	5 (45.5%)	0.016

Variable	Success group n=90	Failure group n=11	p value
After last BPA session			
NYHA Fc (I,II / III,IV) (%)	91.7 / 8.3	44.4 / 55.6	0.002
6 minutes walk distance (m)	442 ± 121	363 ± 106	0.066
mean PAP (mmHg)	29.6 ± 7.5	42.8 ± 8.7	<0.0001
Cardiac Index (L/min/m^2)	3.04 ± 0.70	2.59 ± 0.62	0.047
PVR (wood unit)	3.64 ± 1.61	6.77 ± 2.39	<0.0001
% decrease of PVR (%)	-53.3 ± 19.1	-10.8 ± 9.6	<0.0001
% decrease of mean PAP (%)	-33.2 ± 17.4	-4.5 ± 9.4	<0.0001
Number of patients (mean PAP ≤30mmHg)	53	0	

ICC17-39. IS PRESSURE-WIRE USEFUL TO PREDICT REPERFUSION PULMONARY EDEMA AFTER BALLOON PULMONARY ANGIOPLASTY IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION?

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Aims: To determine whether maintaining mean pulmonary arterial pressure (PAP) distal to target lesions after balloon pulmonary angioplasty (BPA) <35 mmHg protects against reperfusion pulmonary edema (RPE).

Methods: We began BPA in May 2013. Since September 2015 we measured "Pd" value (mean PAP distal to target lesions) after dilation of every branch with pressure-wire, to maintain "Pd" value <35 mmHg.

Results: 156 consecutive BPA sessions in 46 non-surgical patients, 74 BPA sessions without and 82 with pressure-wire guidance, 212 branches measured with pressure-wire. Twenty were unclear lessions with pressure ratio (FFR) >0.80, not treated. The final mean FFR and final mean Pd after BPA in 192 treated lesions were 0.52 and 23 mmHg respectively. Only 13 lesions had final FFR >0,80. The incidence of RPE ≥ grade 2 was similar in the no-guided sessions and in the pressure-wire guided sessions, 5/74 (6,7%) and 4/82 (4,9%) respectively, p not significant. Pd > 35 mmHg after BPA did not correlate with RPE: 3/65 (4,6%) in the sessions with Pd ≤35 mmHg vs 1/11 (9.1%) in the sesions with Pd >35 mmHg (p not significant). According to multivariate analysis the unique factor related to ≥ grade 2 RPE was mean PAP previous to BPA (OR 1.13, CI 1.001-1.282, p=0.04).

Conclusions: The use of pressure-wire to guide BPA did not reduce RPE after BPA. Pd >35 mm Hg distal to target lesions after BPA did not correlate with RPE. Mean PAP before BPA was the only factor related to the appearance of RPE.

ICC17-40. RECURRENCE OF PLUMONARY HYPERTENSION AFTER BALLOON PULMONARY ANGIOPLASTY

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Background: It has been reported that balloon pulmonary angioplasty (BPA) can be an effective therapeutic option for patients of chronic thromboembolic pulmonary hypertension (CTEPH). After the successful BPA, it has been well known to reduce pulmonary arterial pressure (PAP) till almost normal ranges. However, it has not been fully concluded that the long-term beneficial effect of BPA can be observed all of patients with CTEPH.
Objective: The present study was conducted to examine whether BPA chronically maintain PAP within almost normal range in all of patients.
Methods and Results: We conducted BPA in 20 patients with CTEPH, and successful procedure could be obtained all of patients. After the BPA, the significant reduction of PAP was observed all of them. However, after the 6 months of the BPA, the recurrence of pulmonary hypertension was observed in 3 patients (Table). Two patients were asymptomatic and 1 patient suffered from worsening of shortness of breath. In the clinical characteristics, all of cases were women and direct oral anticoagulants (DOAC) were prescribed. Although hemodynamics and symptom were improved immediately after BPAs, hemodynamics were worsened again after 6 months later (Table).
Conclusion: In some of patients with CETPH, BPA may not be sufficient for the long-term outcome of reduction of PAP. Such cases were observed in the patients with prescribing DOAC.

	Case 1	Case 2	Case3
Age	77	79	75
Gender	Female	Female	Female
Treatment	PEA + 4 additional BPAs	4 BPAs	3 BPAs
Change of Symptom before and after treatment	WHO FC 3⇒1	WHO FC 3⇒1	WHO FC 3⇒1
Symptom in chronic period	WHO FC 1	WHO FC 1	WHO FC 2 (worsening)
Mean PAP / PVR / CO before Treatment	46 / 1042 / 2.84	28 / 411 / 2.92	30 / 366 / 3.06
Mean PAP / PVR/ CO after Treatment	22 / 271 / 4.13	15 / 251 / 3.19	25/ 157/ 5.51
Mean PAP / PVR / CO in chronic period	35 / 451 / 3.37	24 / 450 / 3.20	37 / 306 / 4.18
Anticoagulation	Edoxaban 30mg	Rivaroxaban 15mg	Edoxaban 30mg

PAP: pulmonary arterial pressure, PVR: pulmonary vascular resistance,
CO: cardiac output, PEA: pulmonary endarterectomy,
BPA: balloon pulmonary angioplasty,
WHO FC: World health organization functional class

ICC17-41. EVALUATION OF THE ACUTE EFFECT OF BPA BASED ON LESION TYPE USING PRESSURE WIRE IN CTEPH

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Background/Objective: The angiographic types of lesions in chronic thromboembolic hypertension (CTEPH) are classified ring like stenosis, web, abrupt narrowing, complete vascular obstruction (CVO), and pouch. However, few studies have reported on characteristics of pressure gradient and efficacy of balloon pulmonary angioplasty (BPA) in each lesions. Our purpose is to figure out the difference of each lesions in face to BPA for optimizing BPA strategy.

Method: Between October 2012 and January 2017, we evaluated consecutive 218 lesions (32 patients, 88 sessions) measured pressure gradient using pressure wire during BPA at Kobe University Hospital.

Result: Pressure ratio (Pd/Pp: pressure at distal / proximal) in each lesion before and after BPA are shown in the Table. There were 29 lesions detected thrombus by intravascular ultrasound with normal in angiography. The Pressure ratio before BPA were high in order of normal, ring like stenosis, web, abrupt narrowing, CVO. Pressure ratio in each lesion except CVO were significantly improved to similar level in comparison to limited improvement in CVO.

Conclusion: Acute release of pressure gradient by BPA was similarly obtained in all stenotic lesions but not obstructive lesions.

Table

Changes of Pressure ratio in each lesion by BPA

	all	normal	ring like stenosis	web	abrupt narrowing	CVO
n	218	29	72	85	30	2
Pre-BPA	0.48 ± 0.24	0.78 ± 0.14	0.48 ± 0.23	0.45 ± 0.22	0.34 ± 0.18	0.05 ± 0.05
Post-BPA	0.75 ± 0.19	0.86 ± 0.12	0.73 ± 0.20	0.74 ± 0.19	0.76 ± 0.15	0.26 ± 0.11
P value	<0.001	0.001	<0.001	<0.001	<0.001	0.43

normal: lesions detected thrombus by intravascular ultrasound with normal in angiography

CVO: complete vascular obstruction

Medical treatment and interventional treatment (continued)

ICC17-42. REGRESSION OF LEFT MAIN COMPRESSION IN PATIENT WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION AFTER BALLOON PULMONARY ANGIOPLASTY

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Background: Balloon pulmonary angioplasty (BPA) is an effective method reducing pulmonary hypertension (PH) in non-operable CTEPH patients. Due to high PH pulmonary artery dilatation in these patients can cause left main eccentric compression syndrome (LMCS). Incidence of LMCS is unknown but ranging from 5% to 44% in patients with pulmonary hypertension (PH).

Case Report: A 61 year-old woman admitted to PH department with severe dyspnea and 12 years history of CTEPH. According to right heart catheterization, angiopulmonography and coronary angiography the best option for patient was pulmonary thrombendarterectomy (PTE). But due to high pulmonary vascular resistance (PVR) - 20 WU and LMCS – 90% stenosis the operation risk was considered high. Despite proximal type of lesion the multidisciplinary team decided to start treatment with BPA. In 5 months 4 BPA were performed, 10 segmental arteries of both lungs and main lower lobe artery were successfully treated. In a follow-up mean pulmonary artery pressure decreased from 57 to 37 mm Hg, PVR decreased from 20 to 7 WU. Coronary angiography showed no compression of LM with enlargement of lumen area by IVUS from 9 to 12 mm^2. The diameter of PA by CT decreased from 4,2 to 3,6 cm. Patient was transferred to a surgical unit for PTE.

Conclusion: Effective BPA can regression of LMCS in CTEPH patients. BPA can be a preparatory stage in patients with high risk of PTE.

ICC17-43. EFFECT OF BALLOON PULMONARY ANGIOPLASTY ON RIGHT VENTRICULAR AFTERLOAD IN CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Objective: Chronic thromboembolic pulmonary hypertension (CTEPH) results from non-resolving thrombi in pulmonary arteries leading to chronic elevation of right ventricular (RV) afterload. CTEPH is potentially curable by pulmonary endarterectomy (PEA). Balloon pulmonary angioplasty (BPA) is an effective treatment modality for patients who are not suitable for PEA. However, chronic fibrotic material remains in the pulmonary circulation after BPA. Therefore, one might hypothesize that RV afterload remains abnormal after BPA due to increased stiffness of pulmonary vessels. We compared the effects of PEA and BPA on RV afterload in CTEPH.

Methods: Between 4/2014 and 8/2016 17 CTEPH patients (13 inoperable and 4 persistent/recurrent PH after PEA) underwent complete BPA (4.4±2.3 sessions). Hemodynamics were measured at baseline and 6 months after the last BPA session. RV afterload was assessed using standard hemodynamic formulae for pulmonary vascular resistance (PVR) and compliance (C_{PA}). 43 consecutive patients undergoing PEA were used for comparison.

Results: At baseline BPA patients exhibited lower mean pulmonary artery pressure (mPAP; 40.0±12.3mmHg vs 49.2±10.8mmHg, p=0.006), lower PVR (6.0±3.0WU vs 8.4±3.1WU, p=0.009) and higher C_{PA}

(2.6±2.80±3.0mL/mmHg vs. 1.3±0.6mL/mmHg, p=0.011) compared to patients who underwent PEA. However, BPA and PEA led to similar improvements in mPAP (-25.7% [-39.1;-12.6] vs -34.0% [-51.2;-18.2], p=0.252), PVR (-49.4% [-60.4;-31.4] vs. -54.1% [-73.7;-39.8], p=0.564) and C_{PA} (+79.5% [57.3;212.8] vs. +110.7% [38.4;172.5], p=0.554).

Conclusion: Improvement in RV afterload is comparable between BPA and PEA. Our data confirm that BPA is a valuable treatment option for patients with inoperable CTEPH and those with persistent/recurrent PH after PEA.

ICC17-44. EFFICACY OF PERCUTANEOUS TRANSLUMINAL PULMONARY ANGIOPLASTY FOR CHRONIC PULMONARY THROMBOEMBOLISM WITHOUT PULMONARY HYPERTENSION

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Objectives: The purpose of this study was to investigate the effectiveness of PTPA for CTPE without PH on pulmonary hemodynamics, oxygenation and exercise capacity.

Methods: Patients with dyspnea on effort of more than NYHA Ⅱ, less than 25mmHg of mean pulmonary artery pressure (PAP) in right heart catheterization, organized thrombi in pulmonary angiography and segmental multiple defects in VQ scan were defined as CPTE without PH. This study retrospectively included 20 patients with CPTE without PH before the treatment, who underwent PTPA by May 2016. Hemodynamic parameters such as PAP, pulmonary vascular resistance (PVR) and cardiac index (CI), pO_2, and 6-minute-walk distance(6MWD) were compared between before and after PTPA.

Results: The median age, the number of PTPA sessions performed per person and dilated vessels per person were 65[58-73] years old, 2[2-3] and 11.5[8.3-14.5] respectively. PTPA did not cause complications in all patients. Hemodynamic parameters such as PAP and PVR significantly improved after PTPA as compared with the baseline, but CI did not significantly (PAP: 21[20-22] to 17[14-21] mmHg, P<0.01; PVR: 2.8[1.9-3.1] to 1.5[1.2–1.8], P<0.001, CI: 3.2[2.8–3.4] to 3.4[3.0–4.0], P value > 0.05). Oxygenation and 6MWD were improved after PTPA (pO_2 :69.3[66.7–76.1] to 74.7[65.7–89.3] mmHg, P<0.05; 6MWD: 377[333-430] to 401[360-444]m, P<0.05). Ambulatory oxygen therapy (AOT) and PH-targeted drug free rates were reduced significantly (before vs. after; AOT: 80 to 45%, P<0.05; PH-targeted drug free rate: 75 to 25%, P<0.01).

Conclusions: PTPA can safely improve patients with CPTE without PH through oxygenation amelioration.

ICC17-45. THE ROLE OF ECHOCARDIOGRAPHY TO DEMONSTRATE THE EFFECTS OF THE BALLOON PULMONARY ANGIOPLASTY IN PATIENTS WITH CHRONIC THROMBOEMBOLIC HYPERTENSION

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Echocardiography is widely used technique for the assessment of the hemodynamic consequences of pulmonary hypertension (PH). Balloon pulmonary angioplasty (BPA) is an emerging therapeutic method of chronic thromboembolic hypertension (CTEPH).

Aim: To demonstrate the changes of echocardiographic parameters in CTEPH patients before and after BPA.

Material and methods: The study group consisted of 24 patients with CTEPH (age 49-88, mean 72; 12 men). All patients underwent 1-5 BPA sessions. Echocardiography was performed before and after BPA. Seventeen standard echocardiographic parameters and signs useful for assessing the presence of PH were estimated.
Results: Before BPA mean pulmonary artery pressure (mPAP) was 42±7.5mmHg in right heart catheterization. After BPA mPAP decreased to 25.4±6mmHg. We observed echocardiographic improvement of all 17 signs and parameters, 9 of them significant: right and left ventricle diameters ratio (RV/LV): 1.3±0.4 vs 1.1±0.2, p=0.01; inferior vena cava: 21.9±5.6mm vs 17.8±5.6mm, p=0.001; tricuspid annulus plane systolic excursion (TAPSE): 17.8±3.6mm vs 19.9±4.4mm, p=0.02; pulmonary valve flow (PVF) acceleration time: 69±16.2ms vs 79.4±13.9ms, p=0.01; tricuspid regurgitation (TR) peak gradient: 79.9±20.4mmHg vs 62.8±21.5mmHg, p<0.001; RV systolic pressure: 89.9±21.7mmHg vs 70.8±23.6mmHg, p<0.001; the presence: of interventricular septum reverse curvature: 38% vs 17%, p=0.02; of PVF systolic notch: 67% vs 33%, p=0.01; of TR grade moderate or severe: 71% vs 38%, p=0.01.
Conclusion: Echocardiographic signs and parameters revealing right heart hemodynamics improved markedly in CTEPH patients after BPA treatment.

ICC17-46. RESULTS OF BALLOON PULMONARY ANGIOPLASTY

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Introduction: Balloon pulmonary angioplasty (BPA) is a treatment option in chronic thromboembolic pulmonary hypertension (CTEPH). We report results of our 2-year experience.
Aim: To evaluate efficacy and safety of balloon pulmonary angioplasty in patients with CTEPH.
Methods
We enrolled consecutive CTEPH patients disqualified from pulmonary endarterectomy. Treatment goals were improvement in patient's functional capacity to at least class II according to New York Heart Association (NYHA) and decrease in mean pulmonary artery pressure (MPAP). A threshold of MPAP <25mmHg or <30mmHg were used for patients <60 years and >60 years, respectively.
To assess the effect of BPA we measured selected parameters before and 1-month after final BPA as follows: NYHA class (dNYHA), 6-minute walking distance (d6MWD), MPAP (dMPAP), mean right atrial pressure (dRAP), cardiac index (dCI) and pulmonary vascular resistance (dPVR).
Results: We recruited 12 patients (3 males), aged 65 [52–71] years, who underwent a total of 75 BPA sessions (a median of 7 [5;9] for each patient) during which we dilated 431 lesions (a median of 6 [5;7] for each session). We achieved clinical goal in all patients and hemodynamic goal in 7 (58%) patients. Overall we observed improvement in functional capacity (dNYHA= -1 [-1;-1] p=0.002), increase in 6MWD (d6MWD= 80 [50-105]m, p=0.002) and improvement in hemodynamic parameters: dMPAP= -11 [-20;-8]mmHg, p=0.002; dRAP= -3 [-6;0]mmHg, p=0.01; dCI= 0.28 [0.05;0.74] L/min/m^2 p=0.04; dPVR= -2.8 [-5.9;-2.0]WU, p=0.002. None of the patients has died.
Conclusion: BPA is an effective and safe procedure in treatment of chronic thromboembolic pulmonary hypertension.

ICC17-47. CHANGES IN SURFACE ELECTROCARDIOGRAM AFTER HEMODYNAMICALLY EFFECTIVE BALLOON PULMONARY ANGIOPLASTY IN PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Background: We hypothesized that right ventricle (RV) reverse remodeling defined as a changes in electrocardiographic (ECG) markers of RV overload would occur after BPA in patients with chronic thromboembolic pulmonary hypertension (CTEPH).

Methods: In 26 patients with CTEPH (60±21 years old), conventional 12-lead ECG (10mm = 1mV, 25mm/s) was recorded before and after at least 3 BPA sessions.

Results: Twenty-six patients after BPA were divided into 2 groups: group A, defined as a drop in pulmonary vascular resistance (PVR) above the median value for the whole studied population, and group B, defined as a drop in PVR below the median value. The median value of reduction in PVR was 37,94% ($p < 0,001$ between the groups). In group A, there was a significant change after BPA in QRS axis, T axis, P wave amplitude in D II, S wave amplitude in lead V5, R:S wave ratio in lead V5, QRS axis above 110 degrees and the number of patients with S wave amplitude above 300uV in lead V6 ($p<0.05$ respectively) as opposed to group B (Table 1). With respect to percentage improvement in ECG markers of RV overload, only increase in R:S wave ratio in lead V5 significantly correlated with reduction in PVR (%) for the whole studied population ($r= -0,51$, $p<0,05$; Figure 1).

Conclusions: After BPA, a significant improvement in RV overload parameters could be observed by ECG. Whether it has a prognostic significance and can contribute to assessment of success defined with BPA in individual patients requires further studies.

Table 1.

S wave amplitude above 300uV in lead V6	61,54%	38,46%	**p < 0.05**	46,15%	61,54%	**NS**

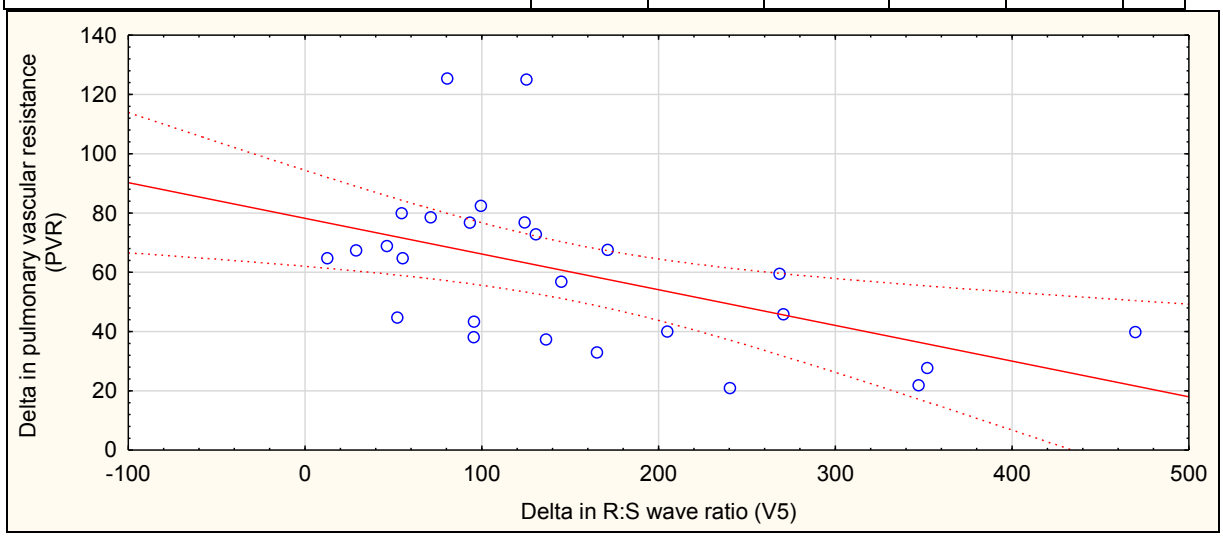

Figure 1.

ICC17-48. REFINED BALLOON PULMONARY ANGIOPLASTY IN PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION - 100 BPA SESSIONS WITHOUT MAJOR COMPLICATIONS IN A SINGLE CENTER

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Objectives: Balloon pulmonary angioplasty (BPA/PTPA) is a novel promising therapeutic method in CTEPH. Our aim was to prove the safety and efficacy of refined BPA/PTPA driven by combined assessment of intra-arterial anatomy (IVUS/OCT) and physiology (pulmonary pressure ratio, PPR) in "all comers" CTEPH pts.

Methods: From July 2014 to March 2017 26 CTEPH pts (mean age 73, 52-88, 13 males) were enrolled to the BPA/PTPA program in our centre according to the following inclusion criteria: (mPAP >30mmHg; WHO class > II). Overall, 26 pts underwent 100 BPA sessions (mean 3 sessions per patient, range 1-12), 245 segmental and subsegmental and lobar pulmonary arteries were dilated (mean 8 vessels per patient, range 1-26). All the angioplasties were performed according to our previously published algorithm (Roik M et al. Int J Cardiol 2016, 203: 228-235).

Results: BPA resulted in clinical and hemodynamic improvement in all enrolled patients (Table 1). All dilated arteries were patent at angiographic reassessment. No significant complications occurred during BPA sessions and follow-up. All treated patients are still alive.

Conclusions: Refined BPA with assessment of intrapulmonary physiology using a pressure wire and precise evaluation of anatomy provides hemodynamic and functional improvement, with minimal complications rate in all-comers patients with CTEPH. This observation requires further validation in a large multicenter prospective study.

TABLE. 1 Clinical, hemodynamic parameters and characteristics of treated lesions and summary of complications in the study group.

	Pre BPA N=26	P	Post BPA* N=18
Mean PAP, mmHg	44 (31-74)	< 0.01	26 (17-33)
WHO functional class 1/2/3/4 (n)	0/0/19/7	<0.05	8/10/0/0
Characteristics of treated lesions and summary of complications			
Total BPA sessions	N = 100		
Number of BPA sessions per patient	3 (1-12)		
Number of dilated vessels in whole group	245		
Number of dilated vessels per patients	8 (1-26)		
Size of balloon used (webs/ring-like stenosis/complete obstruction)	1.0 mm – 9.0 mm		
Reperfusion pulmonary oedema (RPE) (5 grade scale) (%)	1 2 >=3		93 6 1
Arrhythmia within 24 hour after BPA/worsening of renal function within 72 hours after BPA; (n/n)	2 Atrial fibrillation / 2 CIN		
RHF during BPA sessions/ unscheduled hospitalization due to RHF during long term FU/other hospitalizations	0/3/1(COPD exacerbation)		
Deaths (periprocedural or during follow up)	0		

* after final BPA

ICC17-49. BALLOON PULMONARY ANGIOPLASTY - A CURATIVE METHOD IN VERY ELDERLY PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Objectives: Balloon pulmonary angioplasty (BPA) is proposed as a potential therapy of non-operable CTEPH. The aim of the study was to analyze clinical and hemodynamic results of sequential BPA performed in very elderly patients (>75yo) with surgically accessible lesions disqualified from pulmonary endarterectomy (PEA) due to high perioperative risk.

Methods: Ten patients (4 M, 6 F) with median age of 81 years (75 - 88) with confirmed CTEPH (mPAP > 30 mmHg, WHO class > II) were enrolled to BPA program In 7pts lesions were detected at least the level of lobar arteries; while in 3 others lesions were visualized in segmental arteries with no more proximal thrombi.

Results: Overall, 10 pts underwent 39 BPA sessions (mean 3.9 sessions per patient, range 1-9), 70 pulmonary arteries were dilated, (mean 6.5 vessels per patient, range 1-14).

BPAs resulted in a significant clinical and hemodynamic improvement in every patient – Table 1.and resulted in normalization of mPAP (<25mmHg) in 6 of 10pts and mPAP decreased to 25-30 mmHg in 3 others. In the whole group mPAP decreased from 41 (31-53) to 23 (17-33)mmHg (p <0.01). CTEPH patients were followed for a median of 553 days (range 81 -784). No severe complications occurred during BPA therapy and follow-up and all studied patients are still alive and in good general condition.

Conclusions: This study demonstrated safety and efficacy of refined BPA in patients aged at least 75 years with surgically accessible CTEPH disqualified from PEA.

Table 1. Clinical and hemodynamic parameters.

	Pre BPA	P	Post BPA
Mean PAP, mmHg	41 (31-53)	< 0.01	23 (17-33)
CI, l/min/m2	2,31 (1,73 – 2,98)	< 0.01	3,15 (2,38-3,44)
PVR, Wood units	8,21 (4,8-14,38)	<0.01	3,27 (2,1-4,8)
WHO 1/2/3/4 (n)	0/0/7/3	<0.05	2/8/0/0

ICC17-50. FIRST RESULTS OF BALLOON PULMONARY ANGIOPLASTY FOR INOPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH) PATIENTS IN BRAZIL

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Objective: To describe the first results of balloon pulmonary angioplasty (BPA) in Brazil.

Methods: From February 2015 to December 2016, we performed BPA in selected inoperable CTEPH patients in World Health Organization (WHO) functional class III or IV.

Results: Four female patients (mean age, 44,5±9,7 years) with average symptoms duration of 35±16,7 months, underwent BPA. All patients were in continuous use of pulmonary vasodilators. BPA was performed in a step-wise manner (average of 6 procedures for 10±1,6 vessels per patient). After an average of 4,8±1,5 months of follow-up,

mean pulmonary artery pressure (mPAP) decreased 19,4% (63,3±6,4 to 51±3,5 mmHg; p=0,016), pulmonary vascular resistance (PVR) decreased 34% (994±104,8 to 650,8±132 dyne.s.cm^{-5}; p<0,02), and cardiac index increased 18% (2,19±0,5 to 2,6±0,3 l/min/m^2; p=0,15) compared to baseline. Brain natriuretic peptide also decreased (472±357 to 294±310,2 pg/ml; p<0,03). Improvement of subjective symptoms (WHO class III/IV to class II) and quality of life measured by *Minnesota Living with Heart Failure* score (81,7±3,4 to 31±18; p=0,01) were observed. Clinically manifested reperfusion pulmonary edema was diagnosed after 17% of the procedures. Mechanical ventilation was not required. All patients remained alive until the last evaluation.

Conclusions: To our knowledge, this is the first report of a BPA program for inoperable CTEPH patients in our country. All patients presented improvements in hemodynamic parameters, functional status and quality of life. However, persistence of pulmonary hypertension was observed. This may be explained by our BPA learning curve and by the patients extremely high mPAP and PVR baselines levels.

ICC17-51. REFINED BALLOON PULMONARY ANGIOPLASTY FOR THE TREATMENT OF NON-OPERABLE CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION (CTEPH): INITIAL EXPERIENCE IN A CTEPH CENTRE

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Background: The treatment for non-operable chronic thromboembolic pulmonary hypertension (CTEPH) has recently changed with the availability of balloon pulmonary angioplasty (BPA). We started our BPA program in November 2015. In order to minimize the rate of complications, we preferentially target low-risk lesions (ring-like stenoses, webs, and incomplete occlusive lesions in lower lobes) with undersized balloons according to a predefined strategy.

Methods: Between November 2015 and March 2017, 30 patients with non-operable CTEPH were selected in a multidisciplinary meeting. At inclusion, 23 of the 30 patients were already treated with PAH-targeted drugs including riociguat (n=14), endothelin receptor antagonists (n=13), and phosphodiesterase type-5 inhibitors (n=7). A total of 131 sessions have been performed. Fourteen patients underwent complete BPA procedures, and were reevaluated 3 months after the last BPA session. We classified BPA-related complications as: 1) pulmonary injury - defined as hypoxia and clinical symptoms with newly developed radiographic opacities and with or without haemoptysis. - Severity was classified as mild: if patients required nasal oxygen therapy, moderate: required non-invasive ventilation, severe: required mechanical ventilation and/or circulatory assistance. 2) Pulmonary artery dissection

Results:

1/Safety was analyzed in 30 patients and 131 sessions:

Event	Total number	% sessions	% patients	treatment
Pulmonary injury	18	13.7%	36.7%	
Mild	16	12.2%	33.3%	
Moderate	2	1.5%	6.7%	
Severe	0	0.0%	0.0%	
Haemoptysis (+)	16	12.2%	30.0%	
Mechanism of PI				
Wire perforation	10	7.6%	23.3%	Balloon sealing n=10

Balloon injury	5	3.8%	13.3%	Balloon sealing n=5 Embolization n=1
Unknown	3	2.3%	10.0%	Balloon sealing n=1 No treatment n=2
Arterial dissection	3	2.3%	10.0%	No treatment n=3
Death	0	0.0%	0.0%	

2/Efficacy was analysed in 14 patients.

BPA dramatically improved hemodynamic parameters with a significant increase in cardiac output (+ 20%) and a decrease in mPAP (-29%) and pulmonary vascular resistance (PVR) (-41%), which were associated with improved WHO-Fc and 6 MWD

Variables	Baseline	After BPA	P value
CO (L/min)	4.64 ± 0.88	5.57 ± 1.53	0.001
mPAP (mmHg)	44.0 ± 10.5	31.4 ± 10.7	0.035
PVR (dyne/cm^5)	7.72 ± 2.95	4.57 ± 3.09	<0.001
6MWD (m)	403 ± 80	445 ± 99	0.008
SvO2 (%)	63.0 ± 6.1	67.9 ± 5.2	0.003
NYHA Fc (I,II /III, IV)	5 / 9	13 / 1	0.004

Conclusions: Our initial experience with a refined BPA strategy in non-operable patients is encouraging with low rate of complications and significant clinical and hemodynamic improvement.

ICC17-52. HEMODYNAMIC AND CLINICAL IMPROVEMENT AT FOLLOW-UP AFTER SAFE REFINED BALLOON PULMONARY ANGIOPLASTY IN NON SURGICAL PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Aims: To evaluate safety and efficacy of refined balloon pulmonary angioplasty (BPA) in non-operable European population with CTEPH. To determine factors related to reperfusion pulmonary edema (RPE).

Methods: We began refined BPA in inoperable severely ill patients with CTEPH May 2013. We analyzed hemodynamic, functional and biomarkers improvement after ≥ 3 BPA procedures and the incidence of severe complications, RPE and mortality.

Results: We performed 156 BPA sessions in 46 patients. In the 28 patients with ≥ 3 procedures we achived a mean pulmonary vascular resistance (PVR) reduction of 44% (11±5 vs 5.5±2.2 WU p <0.001), a mean reduction in mean pulmonary arterial pressure of 27.9% (53±12 vs 37.8±9.6 mmHg, p <0.001), a mean cardiac index increase of 25.8% (2.3±0.6 vs 2.8±0.6 L/min/m^2), a mean reduction in NT pro-BNP levels of 64.8% (1540± 423 vs. 322.5±397 pg/dL, p <0.001) and a mean increase of 82 meters in the 6-minute walking test (382.8±116 vs. 456±118 m, p=0.003). RPE appeared in 9 procedures (5.8%). One patient had severe RPE that required mechanical ventilation, dying as a consequence of this (mortality 2.1%). By multivariate analysis, value of PVR prior to BPA

procedure and mean wedge pressure prior to BPA correlated with the appearance of RPE (OR 9.92, CI 1.98-49.7, p=0.005 and OR 2.81, CI 1.05-7.56, p= 0.04 respectively).

Conclusions: In our experience, BAP in patients with non-operable HPTEC, performed following a refined procedure, is an effective technique, since it significantly improves hemodynamics, functional capacity and biomarkers, with low ERP and mortality rates.

Baseline characteristics

Patients n	46
Women	32 (69.6%)
Age	59,3 ± 15 years (22-84)
Associated Conditions / Thrombotic Risk Factors	
• Neoplasm	9 (19.6%)
• Thrombophilia	4 (8.7%)
• Central catheters	2 (4.3%)
• Thyroid disease	12 (28.6%)
• Splenectomy	0
HAP specific medication	
• Without drugs	0
• 1 drug	18 (39.1%)
• 2 drugs	21 (45.6%)
• 3 drugs (including prostanoids)	7 (15.2%)

BPA procedure characteristics

Number of lobes treated per session	1.17 ± 0.3 (1-2)
Number of segmental branches treated per session	2.26 ±0.8 (1-5)
Number of subsegmental branches treated per session	3.29 ± 1.6 (1-12)
Mean amount of contrast media used per session	308 ± 80 cc (70-490)
Average procedure lenght	123 ± 25 min (55-205)
Mean fluoroscopy time/procedure	36 ± 11 min (4-61)
Median patient radiation exposure/procedure	86.25 mGy/cm2 (62.3-129.7)
Balloon number/session	2,43 ± 1 (1-5)

BPA procedure complications

Absence of complications	112 (71.8 %)
Guide catheter dissection without loss of flow	10 (6.4%)
Reperfusion pulmonary edema	9 (5.8%)
Perforation requiring interventionism	1 (0.6%)
Hemo sputum	20 (12.8%)
Balloon or guide wire dissection	1 (0.6%)
Femoral hematoma	1 (0.6%)

Changes in hemodynamic parameters, functional class and biomarkers during the follow-up after ≥ 3 BPA

	Before BPA	After ≥ 3 BPA	P
WHO functional class (I / II / III / IV)	3.1 (0/2/19/7)	1.7 (9/18/1/0)	<0.001
6 minutes walking test	382.8 ± 116 m	456 ± 118 m	=0.003
Mean pulmonary arterial pressure	53±12 mm Hg	37.8± 9.6 mm Hg	<0.001
Pulmonary vascular resistance	11±5 W.U.	5.5±2.2 W.U.	<0.001
Cardiac index (L/min /m^2)	2.3±0.6	2.8±0.5	=0.001
Oxygen saturation %	93.9±4.1	96.6±3.3	<0.001

	Before BPA	After ≥ 3 BPA	P
Mixed venous saturation %	61.3±7.4	68.7±5.2	<0.001
NT-proBNP	1540 ± 1423 pg/dl	322.5±397 pg/dl	<0.001

ICC17-53. IMPACT OF THE EFFECT OF ADDITIONAL BALLOON PULMONARY ANGIOPLASTY FOR RESIDUAL PULMONARY HYPERTENSION AFTER PULMONARY ENDARTERECTOMY FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Background: Balloon pulmonary angioplasty (BPA) has been much focused as a treatment procedure for chronic thromboembolic pulmonary hypertension (CTEPH). However, the efficacy of additional BPA for residual pulmonary hypertension (PH) after pulmonary endarterectomy (PEA) has not been fully clarified.

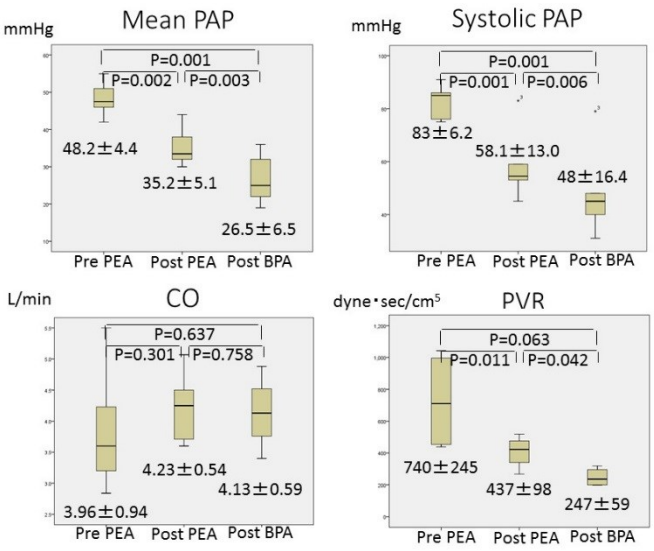
Purpose: The purpose of this study was to evaluate the efficacy of additional BPA for residual PH after PEA for CTEPH.

Methods: Thirty five CTEPH patients were invasively treated in our institute during the past 2 years from January 2015 to December 2016. Fifteen patients received PEA surgically, 9 patients underwent PEA and additional BPA due to residual PH after PEA, 2 patients underwent BPA due to recurrence of PH after PEA in chronic period, and 9 patients were treated by BPA alone. As a prospective observed study, we investigated 7 cases (Male : Female=1:6, Age: 61±16), excluding 2 cases whose treatment was in progress. Hemodynamic values before PEA, after PEA and after BPA were evaluated by right heart catheterization.

Results: Seven patients underwent a total of 25 BPA sessions. A total of 4 (2–6) sessions were performed in each patient, and the number of vessels dilated per session was 5 (1-10). The changes in hemodynamic values were shown in the figure. While cardiac output (CO) was not statistically changed, mean pulmonary artery pressure (PAP) and pulmonary vascular resistance (PVR) were significantly improved.

Conclusion: Additional BPA can be an effective therapeutic option for the treatment of residual PH after PEA.

Figure



Surgical treatment

ICC17-54. PULMONARY ENDARTERECTOMY FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: INITIAL SINGLE INSTITUTION EXPERIENCE

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Methods and Objective: Pulmonary thromboendarterectomy (PTE) is the potential curative treatment for chronic thromboembolic pulmonary hypertension (CTEPH). From January 2014 to December 2016, 10 patients with CTEPH underwent PTE. We analysed clinical characteristics and evolution of these patients.
Results: Median age was 37 (IQR 37-50) years. All patients complained of dyspnea and fatigue. 5 patients had right heart failure. 70% were in NYHA functional class (FC) III-IV and 7 patients were able to perform 6-minute walk test (6MWT) (median 405 meters IQR 345-490). Preoperative mean pulmonary artery pressure (MPAP) 51 mmHg (IQR 31-60) and pulmonary vascular resistance (PVR) 1072 dynas.s.cm^{-5}. (IQR 928-1760). Median time at diagnosis was 13 months (IQR10-19). Cardiopulmonary bypass time 252 minutes (IQR 218-262) and circulatory arrest time 40 minutes (IQR 38-46). Median days of mechanical ventilation 4 (IQR 1-5). After PTE, eight patients normalized MPAP, PVR 440 (IRQ 142-560) dynas.s.cm^{-5} , with significant reduction (p=0.0001). Postoperative complications were pulmonary reperfusion edema and right ventricular failure in 20% and one in-hospital death. ICU stay 8 days (IQR5-12) and the lengh of stay 13 days (IQR8-19). At 12 months (IQR 6-18) of follow up, all patients were in NYHA FC I-II, 6MWT improved in 90 meters (IQR 50-140). Patients enabled to perform the preoperative 6MWT, walked 310 meters (IQR 391-420). Two patients showed persistent pulmonary hypertension; one patient development pulmonary hypertension due to peripheral vasculopathy . All patients were on riociguat.
Conclusions: PTE offers good results for treating CTEHP, improves FC and resolves pulmonary hypertension in most patients.

ICC17-55. CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION - 10 YEARS SINGLE CENTER EXPERIENCE

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Objective: Chronic thromboembolic pulmonary hypertension (CTEPH) is a progressive disease caused by non-resolving but organizing pulmonary thrombo-emboli and vascular remodeling. The epidemiologic and prognostic data of CTEPH in our country was until recently unknown. The aim of the study was to estimate the prevalence and long-term outcome and to evaluate the risk profile of adult population with CTEPH during 10 years at a single (whole-country) center.
Methods: As our institution represents the only center for the CTEPH management in our country, this long-term study, similar to a registry, reflects epidemiologic data of CTEPH in the adult population. In all CTEPH patients, risk factors were analyzed, detailed hematologic analysis included.
Results: Prevalence of CTEPH in adult population was estimated to be 1,67 per 100,000 inhabitants and survival at 1, 3, 5 and 7 years was 95%, 94%, 89 % and 86% respectively. Identified CTEPH risk factors are shown in Table 1. In comparison to healthy controls a significant decrease in platelet count, higher mean platelet volume, higher spontaneous platelet aggregation and increase in von Willebrand factor and fibrinogen levels was revealed.

Conclusions: Our results (epidemiology, long-term outcome, risk factors) in this long-term study in a single (whole-country) center show high similarity with data in international registries.

Table 1. Risk factors (n=81) (in %)

Pulmonary embolism	79
Recurrent PE	28
Idiopathic PE	20
Deep vein thrombosis	59
Thrombolytic therapy	10
Tyreopathy/thyroid replacement therapy	20
Blood type other than “0”	72
Malignity	6
Morbus Crohn/Ulcerative colitis	6
Splenectomy	3
Pacemaker electrode	2

ICC17-56. TEN YEARS EXPERIENCE OF EXTRA-CORPOREAL LIFE SUPPORT AFTER PULMONARY ENDARTERECTOMY

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Objective: To determine the outcomes of patients with chronic thromboembolic pulmonary hypertension who required an extra-corporeal life support (ECLS) after a pulmonary endarterectomy for cardio-respiratory failure.
Methods: We performed a retrospective study of consecutive patients who required an ECLS implantation after pulmonary endarterectomy between 2005 and 2015 at our center. We determined the pre-operative characteristics, intra-operative characteristics, causes aux ECLS requirement, ECLS strategies, management under ECLS and intra-hospital mortality.
Results: During the study period 52 patients required a postoperative ECLS including 43 peripheral and 5 central extra-corporeal membrane oxigenation (ECMO) and 4 pulmonary artery-to-left atrium pumpless membrane (central Novalung ®). The cause of ECLS implantation were pure respiratory failure in 14 cases, mixte respiratory and hemodynamic failures in 15 cases and hemodynamic failure in 23 cases. The median delay of ECLS implantation after surgery was 1 day (extreme values 0 to 24) ; the median ECLS duration was 7.9 days (extreme values 0 to 22 days). Six patients were successfully bridged to heart-lung or double lung transplantation, and 4 died prior organ allocation after failed pulmonary endarterectomy. Among the 42 patients bridged to recovery, 21 (50%) patients were successfuly weaned from ECLS and survived to hospital discharge. Two patients underwent a rescue pulmonary artery angioplasty while under ECLS.
Conclusions: ECLS allows succesfull multimodality salvage strategies in cases of cardiorespiratory failure after a pulmonary endarterectomy.

ICC17-57. PULMONARY ARTERY DENERVATION IN PATIENTS WITH RESIDUAL PULMONARY HYPERTENSION AFTER PULMONARY THROMBOENDARTERECTOMY: ONE-YEAR OUTCOMES

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Objective. To evaluate efficacy and safety of pulmonary artery radiofrequency denervation (PADN) in patients with residual pulmonary hypertension after pulmonary thromboendarterectomy.

Methods. PADN was performed in 20 patients, male 9 (45%), age 48.5 [41;59.5]. Indication for PADN was mean pulmonary artery pressure (PAP) level > 25 mm Hg according to right heart catheterization 12 month after pulmonary thromboendarterectomy. For PADN efficacy and safety assessment we performed V/Q-scanning, echocardiography, right heart catheterization and 6-minute walk test. During preoperative evaluation subsegmental pulmonary artery perfusion defects were revealed in all patients. PADN was performed using nonfluoroscopic 3-D navigation system with standard catheter for radiofrequency ablation. All patients were followed up during 30 days and 12 months after procedure.

Results. There were no complications or death during the procedure and before discharge. Mean procedure time was 105 [93;120] min. After PADN the mean PAP decreased from 37.3 [29;38] mm Hg to 24.6 [17;30] mm Hg **(**p=0.011**)** and pulmonary vascular resistance from 672 [387;566] dyn·sec·cm^{-5} to 386 [155;449] dyn·sec·cm^{-5} (p = 0.017). In 12 months after procedure PAP was 21.3 [15;29] mm Hg (p=0.015) and pulmonary vascular resistance 313 [124;410] dyn·sec·cm^{-5} (p=0.021). 6-minute walk test distance increased from 427 m [352;510] to 492 m [385;675], (p=0.02), and to 512 m [385;701] (p=0.011) in 12 months' follow-up. All patients noticed reduction of dyspnea and improving exercise tolerance.

Conclusions. Immediate and 1-year follow-up results after PADN demonstrated that this procedure in patients with residual pulmonary hypertension after pulmonary thromboendarterctomy is rather safe and effective.

ICC17-58. PERSISTENT EXERCISE LIMITATION (EL) AFTER SUCCESSFUL PULMONARY ENDOARTERECTOMY (PEA): PREVALENCE AND CAUSE

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After successful PEA, the treatment of choice for chronic thromboembolic pulmonary hypertension, patients can continue to suffer from EL, despite normalization of pulmonary vascular resistance.

Objective: The aim of our study was to define the proportion of patients with EL 1 year after PEA and to analyze its determinants with attention to age and clinical-functional abnormalities at rest.

Methods: This is a retrospective evaluation of 202 patients followed up at 12 months. Patients underwent clinical, hemodynamic, echocardiographic, respiratory function evaluations pre- and post-operatively. 52 patients had not exercise testing because of muscular deconditioning or electrocardiography abnormalities. EL was defined as a distance<400 m at Bruce test or a PaO2<60 mmHg.

Results: EL 1 year after PEA was present in 58 patients (39%, 95%CI 31% to 47%). The distance covered during the Bruce test was 201±113 meters in those with EL and 662±177 meters in the others. At multivariable logistic

regression, age (OR 1.07; 95%CI 1.02-1.12), pulmonary arterial compliance (OR 0.10; 95%CI 0.02-0.46), residual volume (OR 1.02; 95%CI 1.00-1.05), lung diffusing capacity for CO (OR 0.96; 95%CI 0.92-0.99) were significantly associated with EL.

Conclusions: A persistent reduced exercise capacity was present in substantial group of patients (almost 40%) despite normalization of pulmonary vascular resistance. The exercise tolerance has a multifactorial etiology involving also respiratory function abnormalities.

The follow up should include exercise testing. Besides cardiac parameters, respiratory function and symptoms can provide useful information also for appropriate therapy.

ICC17-59. ATRIAL ARRHYTHMIA IS COMMON AND ASSOCIATED WITH INCREASED LENGTH OF STAY AFTER PULMONARY THROMBOENDARTERECTOMY

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Objective: Atrial arrhythmias (AA) are common after cardiac surgeries including pulmonary thromboendarterectomy (PTE). Little is published regarding which patients are at the highest risk of developing post-PTE AA.

Methods: We reviewed 521 consecutive patients referred to UCSD for PTE and examined their demographics as well as their baseline pulmonary hemodynamics to determine risk factors for AA.

Results: Overall, 24.2% of patients developed an AA after PTE. Patient who developed AA had a significantly longer ICU length of stay (median: 5 vs. 3 days, p<0.001) and post-operative length of stay (median: 14 vs. 9 days; p<0.001). Patient who developed AA were more frequently male (63.2% male, p=0.003), older (mean age 60.8 vs. 50.7 years, p<0.001), had a prior history of atrial fibrillation (80.2% of those who developed AA) and were more likely to have undergone concomitant CABG (12.7% vs. 6.6%, p=0.028). There was no difference in pre-operative hemodynamics between those who did and did not develop AA. Compared to those who did not develop AA, the cardiopulmonary bypass time was longer among those who did develop AA (261.6 vs. 253.8 minutes, p=0.027). In a multivariate logistic regression model, the pre-operative variables that predicted AA were age (OR 1.058 per year, 95% CI: 1.038-1.078), male sex (OR 1.68, 95% CI: 1.06-2.64), prior AA (OR 2.52, 95% CI: 1.23-5.15) and baseline right atrial pressure (OR 1.039 per mmHg, 95% CI: 1.000-1.079).

Conclusions: Development of atrial arrhythmia is common and associated with longer lengths of stay. Prophylactic anti-arrhythmic medications should be considered in those patients at highest risk for this complication.

ICC17-60. SURVIVAL BENEFIT IMPROVEMENT OF PULMONARY ENDARTERECTOMY WITH EXPERIENCE GAINED IN A NATIONAL EXPERT CENTER

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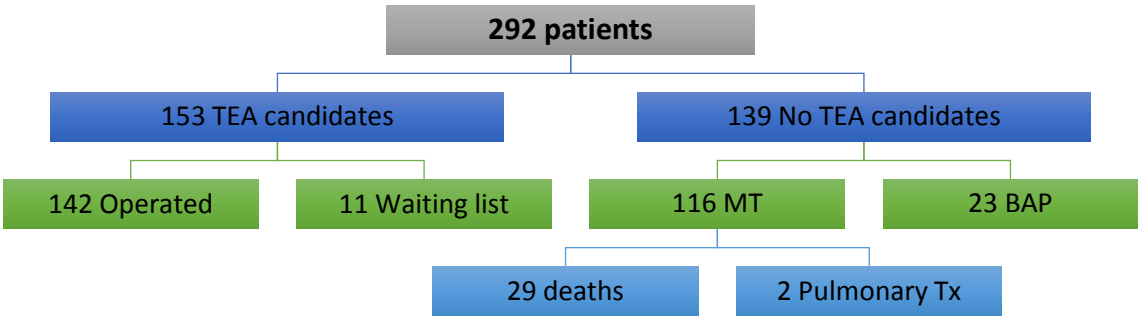
Introduction: Pulmonary Endarterectomy (PEA) is the treatment of choice. Medical Therapy (MT) and Balloon Pulmonary Angioplasty (BAP) should be considered in non-operable patients

Purpose: The aim of this study was to analyse the survival benefit of TEA in an expert center.

Methods: we included all patients evaluated by a CTEPH team from January 1996 to December 2015. Assessment of operability was made by a multidisciplinary team. Learning curve is the period when <10 procedure/year were performed (1996-2010). We analysed survival benefit of TEA vs. MT before/after learning curve. BAP group was excluded.

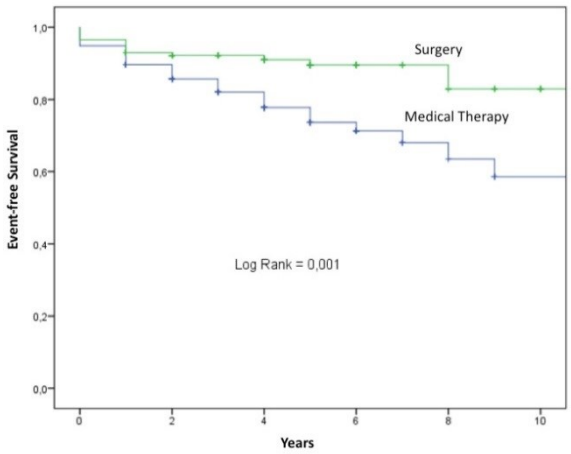
Results: 292 patients were evaluated by a multidisciplinary team. 153 patients (52.4%) were referred to surgery (142 operated before 12/31/2015). Of the 139 non-operable patients, 23 BAP (7.9%) and 116 (39.7%) MT. In the learning curve, 46 were operated and 41 received MT. Table 1 shows baseline differences. In the surgical group, 8 died in the perioperative period and 7 during follow-up. In the medical group, 2 underwent lung transplantation y 29 died. In a multivariate analysis, factors related with mortality were: surgery (RR 0,37; 95 % CI 0,19-0,72), PVR (RR 1,19; 95% CI 1,02-1,39), 6MWT (RR 0,79, 95% CI 0,73-0,85), previous pulmonary embolism (RR 0,50; 95 % CI 0,27-0,92), and cancer (RR 2,57; 95% CI 1,22-5,43). In the survival analysis, we obtained a significant improvement in mortality in the TEA group. This benefit was already present in the learning curve.

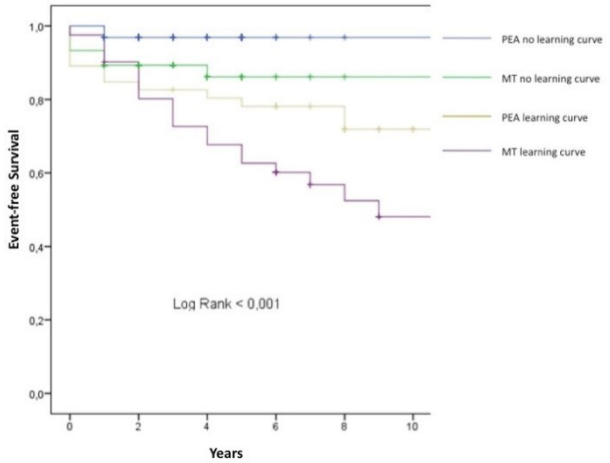
Conclusion: TEA surgery is a safe and effective procedure for CTEPH even in early stages.



	TEA group (n=142)		MT group (n=116)		*p-value*
Variables	**n**		**n**		
Age, years	142	53,62 ± 14,29	116	64,02 ± 15,33	**<0.001**
Gender (male), n (%)	142	80 (56,3)	116	45 (38,8)	**0.005**
BMI, kg/m^2	122	27.03 ± 4,18	83	28,46 ± 4.68	**0.024**
Hypercoagulability, n (%)	134	63 (47)	106	34 (32,1)	**0.019**

		TEA group (n=142)		MT group (n=116)	*p-value*
Antiphospholipid syndrome, n (%)	134	26 (19,4)	106	11 (10,38)	**0,044**
6MWT, m	100	391 ± 117	88	354 ± 117	**0.034**
mPAP, mm Hg,	141	48,62 ± 12,03	111	44,57 ± 11,83	**0,008**
Treatment at dignosis, n (%)	142	86 (61)	116	105 (90)	**<0,001**





ICC17-61. PULMONARY ENDARTERECTOMY IN OCTOGENARIAN POPULATION: SAFETY AND OUTCOMES

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Objective: The pulmonary endarterectomy (PEA) is the treatment of choice for patients with chronic thromboembolic pulmonary hypertension (CTEPH). The number of elderly patients into CTEPH population increased over the last years. In literature, no data are reported regarding PEA in octogenarian patients. Our aim in this study is verify the feasibility, safety and efficacy of PEA into this elderly patient's subgroup.
Methods: At our Center, from April 1994 to August 2016, 734 PEAs were performed, of the overall, in this study, we have considered patients who have at least 1 year follow up (n=635). The population was divided into three group based on patient's age: below 60 years (n=259, group A), between 60 and 79 years (n=352, group B) and over 80 years (n=24, group C). Regression models were used to analyze postoperative results, hospital mortality and long-temp survival, in order to assess the old age sensitivity.
Results : The hemodynamic improvement was similar into the three group: 1 year after PEA, for the group A, B and C, the reduction of the median pulmonary artery pressure was respectively -50% (46±12 to 23±8 mmHg), -

48% (43±12 to 23±8 mmHg) and -41% (42±12 to 24±9 mmHg), the decrease of pulmonary vascular resistances was respectively -72% (903 [628-1163] to 212 [138-304] dyne*sec*cm^{-5}), -67% (847 [595-1129] to 250 [174-332] dyne*sec*cm^{-5}), -65% (877 [679-1257] to 317 [188-503] dyne*sec*cm^{-5}) and the increase of cardiac output was respectively +33% (4±1.4 to 6±1.2 L/min), +22% (3.7±1 to 4.7±1 L/min), +23% (3.3±1 to 4±0.8 L/min). The hospital mortality is slightly greater in octogenarian but not significantly different to group B (total PEA patients= 7%; group A=4%, group B=10%, group C=17%; p= 0,006). The survival at 1 year (excluded hospital mortality) is respectively 100%, 96%% and 95%: group B is not dissimilar to group C (p=0,113).

Conclusion: PEA is safety and efficacy for octogenarian patients. The absence of significantly post-operative differences between group B and C is crucial to confirm the feasibility of PEA for octogenarian population.

ICC17-62. PULMONARY ENDARTERECTOMY WITH PREVIOUS PLASMA EXCHANGE AND IMMUNOGLOBULIN INFUSION IN PATIENT WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION, ANTI-PF4/HEPARIN ANTIBODIES POSITIVITY AND CHRONIC IDIOPATHIC THROMBOCYTOPENIA

V. Grazioli[1], A. Sciortino[1], M. Pin[1], V.N. Merli[1], B. Vanini[1], A. Celentano[1], I. Parisi[1], M. De Amici[2], G. Testa[2], C. Perotti[3], C. Del Fante[3], F. Pulcinelli[4], A.M. D'Armini[1]

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Objective: Chronic idiopathic thrombocytopenia (CIT) and anti-platelet factor 4 (PF4)/heparin antibodies are contraindications to surgery and, in particular, to cardiac surgery considering the high heparin dose used during cardiopulmonary bypass (CPB). The use of plasma exchange (PEX) and immunoglobulins - IgG infusion the days preceding surgery is an excellent solution to allow surgery.

Methods: We presented a 77-year-old woman admitted to our Center for dyspnea and pulmonary hypertension. The medical case history showed a chronic idiopathic thrombocytopenia from the age of 55-year-old and a pulmonary embolism 2 years earlier treated by anticoagulant therapy. After an accurate diagnostic work-up, chronic thromboembolic pulmonary hypertension (CTEPH) with surgical indication was identified: the median pulmonary artery pressure (mPAP) was 32 mmHg, pulmonary vascular resistances (PVR) were 515 dyne*sec*cm^{-5} and the cardiac output (CO) was 3,7 L/min. The laboratory analysis confirmed the presence of thrombocytopenia apparently not related to the contemporary discovering of anti-PF4 antibodies positivity. A serotonin releasing assay confirmed the presence of anti-PF4 antibodies. The therapeutic decision was challenging. After multidisciplinary team discussion, the patient underwent to pulmonary endarterectomy (PEA) with previous PEX and immunoglobulins - IgG infusion.

Results: The post-operative period was uneventful. No allergic reactions were recorded. A corticosteroid therapy was introduced to increase the platelet count until the patient reached the usual values. An important improvement of pulmonary hemodynamic parameters was registered after PEA: the reduction of mPAP and PVR were respectively 22% and 60%; the increasing of CO was 26%. The follow-up at 1 year after surgery confirmed excellent post-operative results.

Conclusions: PEX and immunoglobulins-IgG infusion is a great opportunity for patient with CIT and anti-platelet factor 4 (PF4)/heparin antibodies who require a surgical treatment. The case presented is the first reported in literature for CTEPH patient who need PEA.

ICC17-63. PULMONARY ENDARTERECTOMY CAN BE PERFORMED WITH LOW RATES OF BLOOD TRANSFUSIONS

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Objective: Pulmonary endarterectomy for chronic thromboembolic pulmonary hypertension is a complex procedure performed only in specialized centers. The technique requires prolonged use of cardiopulmonary bypass and frequently employs hypothermic circulatory arrest. Our tertiary care medical center has been committed to blood conservation in cardiac surgery. We sought to determine if our strategies have been effective in patients undergoing pulmonary endarterectomy.
Methods: We retrospectively reviewed our institutional experience by analyzing blood transfusions administered both intraoperatively and in the post-operative phase. From March, 2011 to March, 2017 pulmonary endarterectomy was performed on 63 patients at our medical center. All patients underwent retrograde autologous priming of the cardiopulmonary bypass circuit as well as autologous normovolemic hemodilution when tolerated.
Results: Pre-operative mean systolic pulmonary artery pressures were 75 mmHg and postoperatively were 45 mmHg. Operative mortality was 4.7%. At any point during the hospitalization, only 16/63 (25.4%) of patients received allogeneic packed red cell transfusions (9 intraop, 6 postop, and 1 both intraop and postop). Three patients (4.7%) received platelet transfusions and 4 (6.3%) received fresh frozen plasma.
Conclusions: With effective blood conservation strategies, pulmonary endarterectomy can be performed with infrequent need for preoperative blood transfusions.

ICC17-64. MINIMALLY INVASIVE BILATERAL PULMONARY THROMBOENDARTERECTOMY: A NOVEL TECHNIQUE

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Introduction: Pulmonary thromboendarterectomy (PTE/PEA) is a complex surgical procedure requiring median sternotomy, cardiopulmonary bypass with profound hypothermia, and circulatory arrest for a complete endarterectomy. With recent advancements in minimally invasive cardiac surgery, we sought to determine if these techniques could be applied to PTE surgery, avoiding a sternotomy.
Methods/Technique: Complete endarterectomy with resection of segmental and subsegmental disease were performed via mini-anterior thoracotomies, with use of profound hyperthermia and circulatory arrest; after feasibility and exposure were explored and proven using cadaver models. Minimally invasive and percutaneous cannulation techniques were utilized for initiation of bypass, profound hypothermia and circulatory arrest. The usual protocol for circulatory arrest and exposure of the pulmonary arteries were used. A complete endarterectomy was performed with visualization equivalent to that of a sternotomy.
Results: Since the completion of laboratory work, we have offered minimally invasive PTE to two patients. Both patients had excellent outcomes without significant intra-op or post-op complications.
Their results are summarized in Table 1.

	Patient 1	Patient 2
History	39 y/o F, exercise induced dyspnea with complete occlusion RML/RLL	27 y/o F, history of LE DVT, complete occlusion of LPA
Circulatory arrest times	Right: 9min Left: 4min	Right:13min Left-:15min
UCSD Classification Disease Level	Right- level 2 Left- level 0	Right- level 3 Left- level 1C Thickened left PA, small luminal size with evidence of prior vascular inflammation.
PVR dynes.s.cm-5	Pre- 173 Post - 116	Pre- 386 Post- 196
Postop LOS	8 days	12 days
Complications	none	Prolonged LOS due to persistent chest tube drainage
Outcome	Improved hemodynamics and perfusion on V/Q scan	Improved hemodynamics and perfusion on V/Q scan

Conclusion: Though minimally invasive PTE surgery remains in its infancy, we report that our laboratory work and first two patients' results are successful. We have established that the minimally invasive technique is safe and provides equal visualization as compared to traditional sternotomy approach.
Given our limited experience, we are judicious in our patient selection– and cautious about reaching conclusions compared to conventional PTE beyond the feasibility offered by these two cases. With our growing experience, we believe the procedure may offer an alternative and possibly superior option to conventional PTE through median sternotomy.

ICC17-65. IMPROVED SURGICAL OUTCOMES OF DISTAL TYPE CTEPH

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Introduction: Nearly a half of CTEPH patients were not indicated to pulmonary endarterectomy (PEA) mainly due to distal disease. With increased experience, we have applied PEA to distal type CTEPH, and sought to determine surgical outcomes of patients with distal disease in the last 5 years compared with those treated in the previous period.
Methods: We diagnosed 144 patients as distal disease between 1995 and 2015. The extent of central disease quantitated by adding up the number of abnormal central artery up to maximum score of 4 and distal disease was defined as the score of $\leq$1. In the previous period, 30 of 111 patients (27%) underwent PEA (group 1). In the recent period, 20 of 33 patients (61%) and additional 7 patients deemed inoperable due to distal disease in the previous period underwent PEA (group 2).
Results: In-hospital mortality was significantly decreased from 7 patients (23%) in group 1 to 1 patient (3.7%) in group 2 (p=0.02). Postoperative mPAP and PVR of survivors were significantly decreased with no differences between both groups (mPAP: 29±9 vs 26±9 mmHg, p=0.36 and PVR: 401±271 vs 325±178 dyn.s.cm-5, p=0.25). Despite preoperative diagnosis of distal disease, surgical specimens showed that most patients had central disease.

Conclusion: Surgical outcomes of patients with distal disease have improved. Preoperative diagnosis of distal disease may not be definitive, and most patients with distal disease have central disease and can benefit from PEA.

ICC17-66. PERIOPERATIVE EXTRACORPOREAL MEMBRANE OXYGENATION-BASED PROTOCOL FOR ACUTE PULMONARY EMBOLECTOMY: TECHNIQUE AND PRELIMINARY RESULTS

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Objective: At our institution, since 2012, we have applied a protocol using perioperative veno-arterial extracorporeal membrane oxygenation (v-a ECMO) support in all patients with acute pulmonary embolism, in order to reduce the impact of hemodynamic instability. In this study, we present the preliminary results of perioperative ECMO-therapy.

Methods: We retrospectively reviewed all patients who underwent embolectomy due to acute pulmonary embolism at our institution between 11/2012 and 09/2016. The first line therapy was v-a ECMO implanted percutaneously in the femoral vessels. This was intraoperatively switched to cardiopulmonary bypass and then again to v-a ECMO at the end of surgery. V-a ECMO was continued postoperatively at the intensive care unit.

Results: Eight patients underwent pulmonary embolectomy for acute embolism. Preoperatively, 2 (25%) patients showed concomitant chronic thromboembolic pulmonary artery hypertension, 5 (62%) patients required mechanical ventilation, 6 (75%) patents had required v-a ECMO support for cardiogenic shock. V-a ECMO was implanted before the anaesthesia in 2 (25%) patients. Intraoperatively, cardiopulmonary and cross clamp times (minutes) amounted to 135±57 and 66±34, respectively. Six (75%) patients required deep hypothermic circulatory arrest (15±12 minutes). Three (37%) patients underwent combined cardiac procedures. V-a ECMO was continued postoperatively in all patients and weaned successfully after a mean of 4±2 days. Four (50%) patients were successfully extubated before ECMO weaning. Two (25%) patients required rethoracotomy for bleeding, without in-hospital mortality.

Conclusions: Our v-a ECMO based protocol for acute pulmonary embolectomy led to stabilization of preoperative compromised haemodynamic. Postoperative ECMO provided good postoperative results in high risk patients.

ICC17-67. EXTRACORPOREAL MEMBRANE OXYGENATION SUPPORT AFTER PULMONARY ENDARTERECTOMY

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Objective: Pulmonary endarterectomy (PEA) for CTEPH can provide remarkable improvement in pulmonary hemodynamics. However, some patients develop severe cardiorespiratory failure due to reperfusion pulmonary edema, pulmonary hemorrhage, or residual pulmonary hypertension, and extracorporeal membrane oxygenation (ECMO) can be a bridging treatment to recovery. We reviewed outcomes of patients who needed ECMO after PEA.

Patients: Among 63 patients who underwent PEA between Dec/2011 and Jan/2017, 11 patients (17%) needed ECMO after PEA. The indications of ECMO were as follows: residual PH: 8 patients, pulmonary hemorrhage: 2

patients, and right heart failure due to severe TR associated with pacemaker lead. ECMO flow was adjusted allowing right heart ejection to prevent pulmonary thrombosis. Initial ECMO flow was 1.9 (1.0-4.0) L/min, and pulmonary pulse pressure was 11±6 mmHg.

Results: Preoperative mean PAP was 49±7 mmHg, and PVR was 936±439 dyne.s.cm^{-5}. Preoperative ECMO was instituted in 3 patients with acute exacerbation of CTEPH. Central ECMO was instituted in 6 patients and peripheral ECMO through the femoral vessels was in 5. Seven patients (63%) weaned from ECMO with mean duration of ECMO support of 2.3 days, while 2 patients were re-instituted due to cardiorespiratory failure. There were 4 deaths (36%) from recurrent pulmonary embolism in 2 patients, hypoxic encephalopathy in 1, and severe respiratory failure in 1. All of patients who received central ECMO developed cardiac tamponade due to mediastinal hematoma.

Conclusion: ECMO is a useful treatment for patients with cardiorespiratory failure after PEA, bridging them to recovery.

ICC17-68. STAGED HYBRID TREATMENT OF PEA AND BPA FOR CTEPH

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Objectives: The patient characteristics and outcome of a sort of staged hybrid therapy of PEA and BPA for CTEPH are investigated.

Methods: From 2012 to 2017, 53 patients of proximal type in 51 and distal type in 2 underwent PEA for CTEPH. One patient with proximal type resulting in hospital death was excluded from this study. BPA was performed for 13 patients (hybrid group) for residual PH (mPAP> 25 - 30 mmHg) after PEA. The clinical characteristics, hemodynamic data, and operative data were compared between the hybrid (n= 13) and non-hybrid (n= 39) groups.

Results: There were no significant differences in the age and gender. In the hybrid group, the duration of diagnosis to operation was longer; 74±72:37±44 months (p= 0.03). The preoperative PH was also severer: 45.1±7.2:38.8±9.6 mmHg of mean PAP (p=0.04) and 973±440 and 659±359 dyne・sec・cm^{-5} of PVR (p= 0.01). There were no significant differences in the surgical parameters during PEA and the CTEPH lesion sites. The postoperative mean PAP and PVR were significantly higher in the hybrid group; 31.0±8.5:18.1±6.2 mmHg (p< 0.001), 422±183:226±157 dyne・sec・cm^{-5} (p< 0.001). However, through staged BPA, the mean PAP and PVR were decreased favorably to 25.4±7.9 mmHg and 343±200 dyne・sec・cm^{-5} in the hybrid group.

Conclusions: A staged hybrid therapy of PEA and BPA is a promising therapeutic option for surgically difficult patients having distal dominant or combined proximal and distal CTEPH lesions.

ICC17-69. OUTCOME AFTER PULMONARY ENDARTERECTOMY FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION – A GERMAN SINGLE-CENTRE TWO-YEAR EXPERIENCE

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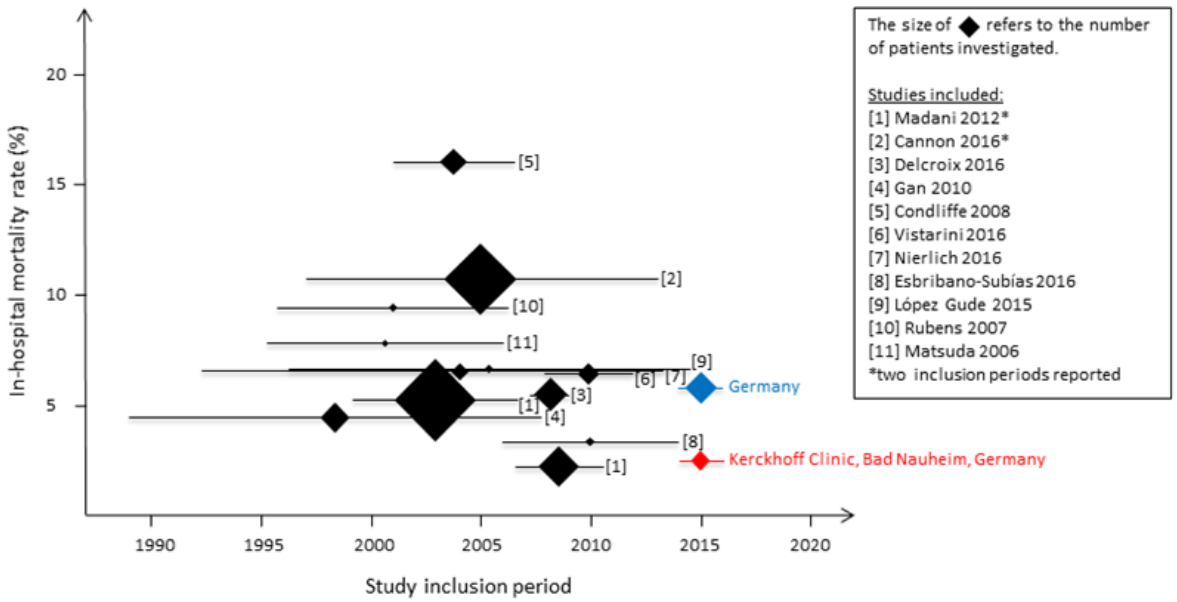
Objective: We investigated the management and outcome of patients with operable chronic thromboembolic pulmonary hypertension (CTEPH) who underwent pulmonary endarterectomy (PEA) at a large German referral centre.

Methods and Results: In Germany, 394 PEAs were performed in 2014 and 2015 with an in-hospital mortality rate of 5.8%. Of these, 253 patients (64.2%) were treated at the Kerckhoff Clinic, Bad Nauheim and 237 (93.7%; median age, 62 [IQR, 52-72] years; 46.0% female) included in the present study. PEA was successful in 236 (99.6%) patients, with reduction of median pulmonary vascular resistance from 7.2 (IQR, 5.0-10.4; n=197) to 4.8 (IQR, 3.5-6.5; n=163; p<0.001) WU and median mean pulmonary artery pressure from 43 (IQR, 34-50; n=206) to 29 (IQR, 25-33; n=171; p<0.001) mmHg. The median duration of surgery was 397 (IQR, 363-431) min. Periprocedural (0%) and in-hospital (2.5%) mortality rates were excellent and lower than previously reported (**Figure 1**). Forty-two patients (17.7%) had intraoperative and 60 (25.3%) postoperative complications. Duration of surgery was the only predictor of in-hospital mortality (≥500 min; OR, 32.0 [95%CI, 5.5-187.6]) and the only independent predictor of both intraoperative (≥440 min; OR, 10.8 [95%CI, 4.4-26.5]) and postoperative (≥390 min; OR, 2.4 [95%CI, 1.1-5.7]) complications. Only intraoperative complications independently predicted a longer duration of surgery (≥397 min; OR, 5.0 [95%CI, 2.2-11.2]).

Conclusions: In an experienced centre with multidisciplinary diagnostic and therapeutic approaches, PEA is safe. Prognosis is mainly determined by the occurrence of intraoperative complications and the duration of surgery.

Figure 1. In-hospital mortality rate after PEA



ICC17-70. INDICATIONS AND OUTCOME AFTER PULMONARY ENDARTERECTOMY WITH OTHER CARDIAC OPERATIONS

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Objectives: Pulmonary artery endarterectomy (PEA) is established as a successful method for the treatment of chronic thromboembolic pulmonary hypertension (CTEPH). A significant fraction of patients indicated for the pulmonary endarterectomy has other severe comorbidities that generally increase the risk of cardiac surgery. The aim of this study is to analyze the process of indications and therapeutic procedures at our centre.
Methods: 291 patients with CTEPH underwent PEA using cardio-pulmonary bypass and deep hypothermic circulatory arrest form 2004 to 2016. 195 patients underwent simple PEA (group A) and 96 PEA with other cardiac operations (group B). In group B 96 patients carry out 109 operations , 42 patients had foramen ovale patent (FOP)closure ,34 patients underwent PEA in conjunction with coronary bypass grafting , 18 patients MAZE procedure with PEA, 8 patients had valve surgery with PEA and 7 patients pacemaker. Statistical analysis with Kaplan –Meier cumulative survival was performed and groups were compared using Cox regression analysis.
Results: Cumulative survival after one year was in group A 94.7%, and B 90.1%. After five years was survival A 83.3% and B 79.3%. After seven years was cumulative survival in group A and in group B 78.9% Only in specific group PEA+ CABG was significantly different from the reference group A (p=0,031).
Conclusions: Cumulative survival in patients after PEA with combine procedure is comparable with patients with only simple PEA procedure and it is satisfactory and very good.
Patients with concomitant myocardial revascularization had statistically significant worst long term cumulative survival.

ICC17-71. PULMONARY SCINTIGRAPHY WITH SPECT-CT TECHNIC FOR THE EVALUATION OF POST OPERATIVE PERFUSIONAL RESULTS IN PULMONARY ENDARTERECTOMY SURGERY FOR CTEPH

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Introduction: Chronic thromboembolic pulmonary hypertension (CTEPH) is one of the potentially curable causes of pulmonary hypertension and is definitively treated with pulmonary thromboendartectomy. Several methods for evaluation exist for diagnostic purposes, but the ventilation/perfusion scan is still the most sensitive, and surgical results are directly linked to the capability to promote reperfusion at all lung segments previously occluded by chronic clots,5 reinforcing the need for a good evaluation of proper disease distribution.
Objectives: evaluate pulmonary scintigraphy with Spect-CT technic as a tool to evaluate surgical success in reperfunding previously obstructed lung segments in CTEPH patients submitted to pulmonary endarterectomy surgery.
Material and methods: We use SPECT-CT scintigraphy in our routine after surgery, patients were studied after 90 days of surgery if clinically stable, and the images are then compared.
We then compare the images with the aim of evaluating restoration of blood in the target areas and possible causes of persistent perfusion defects that could be improved on new cases.

Results: it is possible to see the good correlation of perfusion improvement and anatomic distribution *versus* surgical assessment of the clots and areas approached.
The marked improvement on image exams are corroborated by the good clinical outcome in the moment of the exams, with patients with great improvement in exercise capacity, showing that SPECT-CT can be used as a good and easy to do procedure to evaluate surgical results in CTEPH patients.

ICC17-72. PAVIA EXPERIENCE IN REOPERATIVE PULMONARY ENDARTERECTOMY

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Objective: In our experience we re-performed pulmonary endarterectomy (PEA) in 10 patients who previously underwent a first PEA. We analyzed this cohort of patients in order to investigate the main causes of recurrence of symptomatic pathology and the clinical and hemodynamic results of redo surgery.
Methods: Between 1994 and February 2017, 10 of 771 patients were re-operated at our institution. Postoperative data available were analyzed and a comparison between first and second PEA hemodynamic and clinical results was operated. In-hospital mortality rate was also evaluated.
Results: After re-operation mean pulmonary arterial pressure (mPAP) decreased from 45±9 to 34±10 mmHg and pulmonary vascular resistance (PVR) reduced from 932±346 dyne*sec*cm^{-5} to 428±207 dyne*sec*cm^{-5}. Hemodynamic data revealed redo PEA worthy results, although they are less important than after first PEA. WHO functional class improvement demonstrated satisfactory clinical results. In-hospital mortality of repeat PEA is 40%.
Conclusions: Reoperative PEA operative candidacy should be assessed in case of young patients, no other risk factor and recent medical history of pulmonary hypertension. In the other cases, in-hospital mortality results too high and pulmonary hypertension specific drug therapy or interventional approach should be previous considered.

ICC17-73. OUR EXPERIENCE IN THE MANAGEMENT OF PULMONARY HEMORRHAGE DURING PULMONARY THROMBO-ENDARTERECTOMY

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Introduction: Pulmonary hemorrhage occurring during Pulmonary thrombo-endarterectomy (PTE) is a dreaded surgical complication with a high mortality. The lungs get flooded from the pulmonary artery (PA) inflow when circulation is resumed, and the resulting exsanguination and hypoxia makes separation from cardio-pulmonary bypass nearly impossible. Although rare, it is potentially fatal and the treatments described are only supportive. In this report, we describe the technique to diagnose this breach during surgery and how we applied the method in six cases to effectively treat pulmonary hemorrhage.
Method: In a five-year period from 2011 to 2016, 229 PTEs were performed in our center with injury to the PA wall occurring in 6 cases. Previously, the breach went undetected while the PA was open and empty during deep hypothermic circulatory arrest (DHCA). Since adopting this technique, after the endarterectomy is completed, PA

is flooded with saline and positive airway pressure is instituted using the ventilator circuit. Bubbling of air leaking from the alveoli is seen at the site where the barrier is breached. While still under DHCA, the affected segmental artery is plugged using a wick of Surgicel (Ethicon LLC San Lorenzo, Puerto Rico-00754) and the seal checked again under water against positive airway pressure. We used this technique in all 6 patients and in them, bleeding into the airway was scanty, and stopped completely after Protamine administration.

Conclusion: This technique of detecting and treating PA wall rent before closure and recirculation has significantly improved outcomes after PTE complicated by pulmonary hemorrhage.

ICC17-74. AGE SHOULD NOT BE A BARRIER TO PULMONARY ENDARTERECTOMY IN CAREFULLY SELECTED PATIENTS

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Objective: We previously reported that hospital survival in patients over 70 years undergoing pulmonary endarterectomy (PEA) was comparable to those under 70, albeit with longer hospital and intensive care unit (ICU) stays. Octogenarians have acceptable morbidity and mortality outcomes for selected cardiac surgery. We aimed to assess if this also applies to PEA surgery.

Methods: Consecutive patients undergoing PEA for chronic thromboembolic pulmonary hypertension from June 2006 to August 2016 at Papworth Hospital, UK, were included in this retrospective analysis. The cohort was dichotomized into over and under 80 years according to age at surgery. Pre-operative baseline and post-operative 3-6-month follow-up data were recorded.

Results: 1152 individuals underwent PEA (1115 under 80 years, 37 over 80) during the study period. Baseline and 3-6 month follow up variables and outcomes are summarized in Table 1. Survival is lower in the over 80 group (Log-rank test, P=0.002), but is no different from an age and sex matched UK population (P=0.5) (Office of National Statistics). The hospital length of stay was longer in those over 80 (median, 18 vs. 14days; P=0.001), however, there was no difference in NYHA class, haemodynamics, type of surgical disease, CAMPHOR score or ICU length of stay between the two age groups.

Conclusions: We found similar outcomes in patients under and over 80 undergoing PEA except for a prolonged hospital length of stay and reduced longer-term survival, although survival is comparable to the age-matched population. This suggests that age alone should not be a contraindication for PEA.

	Baseline			Follow-up		
	Under 80	Over 80	P-value[a]	Under 80	Over 80	P-value[a]
n	1115	37				
Age [median (IQR)]	61 (22)	81 (2)				
Gender [female (%)]	46	46	NS			
NYHA class [1/2/3/4 (%)][b]	0 / 14/ 73/ 13	0/ 3/ 70/ 26	NS	28/ 47/ 23/ 1	22/ 40/ 33/ 4	NS
6mwd [metres; median (IQR)]	295 (190)	227 (229)	NS	363 (157)	284(101)	**0.001**
mPAP [mmHg; median (IQR)]	45 (15)	42 (12)	NS	25 (13)	28 (12)	NS
PVR [dynes.s.cm^{-5}; median (IQR)]	666 (479)	743 (462)	NS	235 (212)	297 (294)	NS
CI [L/min/m^2; median (IQR)]	2.18 (0.84)	2.06 (1.05)	NS	2.28 (0.69)	2.26 (0.72)	NS
CAMPHOR: [median (IQR)]						
Symptoms	13 (10)	12 (9)	NS	4 (8)	4 (7)	NS
Activity	11 (9)	14 (8)	NS	6 (9)	9 (6)	NS
Quality of life	11 (11)	11 (11)	NS	4 (10)	6 (8)	NS
Type of surgical disease[c] [1/2/3/4 (%)]	14/ 58/ 26/ 3	14/ 59/ 27/ 0	NS			
Bypass time [min; median (IQR)]	323 (65)	305 (51)	**0.002**			
Arrest time [min; median (IQR)]	37 (15)	33 (13)	NS			
Length of stay [days; median (IQR)]						
Intensive care unit	4 (3)	5 (4.5)	NS			
Total hospital	14 (10)	19 (7)	**0.001**			
In-hospital mortality [n (%)]	45 (4)	3 (8)	NS			
Survival [%]						
1 year				91.8	83.5	**0.002**
3 years				88.2	76.4	
5 years				84.4	69.4	

Table 1. Baseline and follow-up variables and outcomes subdivided by age cohorts. The follow-up time for NYHA, 6mwt, haemodynamics and CAMPHOR score was 3-6 months. NYHA (New York Heart Association), 6mwt (6 minute walking test), mPAP (mean pulmonary arterial pressure), PVR (pulmonary vascular resistance), CI (cardiac index), CAMPHOR (Cambridge Pulmonary Hypertension Outcome Review score), IQR (interquartile range).
[a] P-values were calculated using Chi-squared test or Fisher's exact test for categorical data, Wilcoxon rank-sum test for continuous data and Log-rank test for survival data. A Bonferoni adjusted significance threshold of <0.002 was used. Significant P-values are highlighted in bold. NS (Not significant).
[b] Total percentage may not add up to 100 due to rounding
[c] Jamieson classification.

ICC17-75. ECMO TO TREAT PULMONARY MASSIVE BLEEDING DURING PULMONARY ENDARTERECTOMY

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Objective: during endarterectomy surgery two kinds of pulmonary bleeding may occur: from the disruption of vascular wall with communication between airways and circulation and from systemic-bronchial collateral artery. In the first case bleeding can be controlled by the surgeon inserting BioGlue in the culprit vessel (in the worst cases lobectomy may be needed); in the second case reducing systemic anticoagulation is the only option.
Methods: woman with post-embolic pulmonary hypertension and history of hemoptysis underwent bilateral endarterectomy. During rewarming, broncoscopy (FBS) showed a massive pulmonary bleeding. The patient was cooled down again, extracorporeal circulation stopped and an accurate toilette FBS was done. When flow was re-established, the point of bleeding was located in the medium lobe. During surgery some material was removed from that lobe and the bubbles test was negative. Pre-operative CT scan showed many bronchial-systemic

collateral arteries, so the bleeding should have come from there: no glue was inserted in distal pulmonary branches and no lobectomy was taken in account. We switched to central VA-ECMO and heparin was antagonized. After reaching a normal Activated Clotting Time (132 sec), the bleeding was still present but less important and a bronchial blocker was inserted. The patient was transfused with fresh frozen plasma.
Results: She remained overnight with VA-ECMO with a minimal heparin infusion. In the morning the blocker was removed and no bleeding was seen, then she was weaned from ECMO.
Conclusions: VA-ECMO is a suitable method to treat intraoperatively acute no surgical airways bleeding during pulmonary endarterectomy.

ICC17-76. DYNAMIC VASCULAR CHANGES IN CTEPH AFTER PEA

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Introduction: Recurrent pulmonary embolism has been described after pulmonary endarterectomy (PEA), despite lifelong anticoagulation. However, structured follow-up with CT-pulmonary angiography (CTPA) after PEA was never performed.
Objective: To describe the occurrence of new intravascular abnormalities in the pulmonary arteries 6 months post-PEA in CTEPH and to determine predisposing factors and influence on hemodynamic/functional outcome.
Methods: In this observational analysis ECG-triggered CTPA 6 months post-PEA were compared to CTPA pre-PEA, and scored for new (*i.e.* not present before PEA) vascular lesions (thrombus/webs, collapse/early tapering). Patients with or without new lesions were compared regarding baseline characteristics, hemodynamics and functional outcome at 6-month follow-up, time to adequate anticoagulation post-PEA, type of continued anticoagulant.
Results: 33 CTEPH patients had CTPA performed both before and 6 months post-PEA. 10/33 patients had new lesions 6 months post-PEA, mainly localized in the segmental arteries. Baseline characteristics were only different regarding age. Comparison of the presence of thrombophilia factors was hampered due to incomplete data. Hemodynamics at 6 months were not different except for a higher (but normal) PVR in the group with new vascular lesions. Also anticoagulation parameters did not differ between groups.
Conclusion: In this observational study, CTPA showed new vascular lesions at 6 months post-PEA in 30% of patients. Neither predisposing factors nor clinically significant differences regarding hemodynamic/functional outcomes were found. Possible explanations for our findings include *in situ* thrombosis, local scarring and redistribution of perfusion. Long-term outcome of these changes is currently unknown. However, we have shown that dynamic vascular changes occur after PEA in a significant proportion of patients.

Table: comparison of baseline characteristics, anticoagulation, hemodynamics and functional outcome.

Variable	Group 1: new vascular lesions on CTPA 6 months post-PEA 10 patients	Group 2: no new vascular lesions on CTPA 6 months post-PEA 23 patients	P-value
Age at PEA (years)	68 (62-70)	60 (52-66)	0.041
Male gender	6 (60%)	15 (65%)	>0.999
BMI at baseline (kg/m2)	26.1 (24.2-28.2)	26.4 (24.2-30.2)	0.916
Acute VTE in previous history	4 (40%)	13 (65%)	0.255
Blood group non-O	5 (50%)	18 (78%)	0.215
PAH-specific medication pre-PEA	4 (40%)	7 (30%)	0.696
VCI-filter pre- and post-PEA in situ	8 (80%)	21 (91.3%)	0.567
mPAP pre-PEA (mmHg)	37 (33-45)	41 (35-48)	0.255
PVR pre-PEA (dynes.s.cm-5)	505 (348-665)	462 (329-726)	>0.999
CI pre-PEA (L/min/m2)	2.8 (1.8-3.2)	2.4 (2.1-2.8)	0.933
Time to start heparin after ICU admission (hours)	3.4 (3.1-3.6)	4.1 (3.2-6.1)	0.578
Time to first adequate APTT after ICU admission (hours)	10.9 (9.0-29.0)	12.1 (10.2-28.2)	0.451
Anticoagulation post-PEA with VKA (compared to DOAC)	10 (100%)	21 (91.3%)	>0.999
mPAP at 6 months post-PEA (mmHg)	23 (19-30)	23 (18-27)	0.616
PAWP at 6 months post-PEA (mmHg)	9 (5-12)	11 (7.5-12)	0.603
PVR at 6 months post-PEA (dynes.s.cm-5)	227 (162-272)	160 (90-206)	0.039
CI at 6 months post-PEA (L/min/m2)	2.9 (2.5-3.3)	3.3 (2.7-3.7)	0.144
NT-proBNP at 6 months post-PEA (ng/L)	226 (116-660)	202 (98-364)	0.349
6MWD at 6 months post-PEA (m)	488 (395-566)	494 (456-542)	0.849

Data presented as median (IQR) or number of patients (%). BMI: body mass index; VTE: venous thromboembolism; PAH: pulmonary arterial hypertension; VCI: vena cava inferior; VKA: vitamin K antagonist; DOAC: direct oral anticoagulant; mPAP: mean pulmonary arterial pressure; PAWP: pulmonary artery wedge pressure; PVR: pulmonary vascular resistance; CI: cardiac index; 6MWD: 6 minute walking distance. Statistical tests: Mann-Whitney test (numeric variables) and Fisher's exact test (categorical variables).

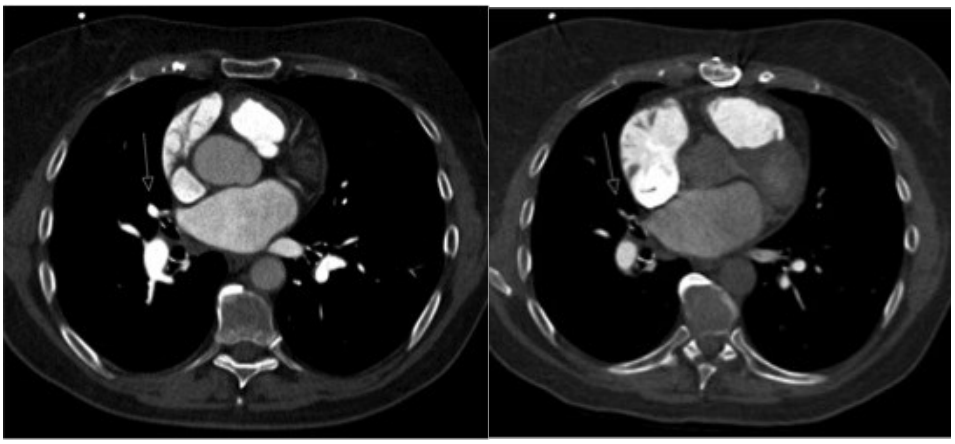

Figure 1: open lateral segmental artery of the middle lobe pre-PEA (left image), occluded lateral segmental artery of the middle lobe post-PEA (right image).

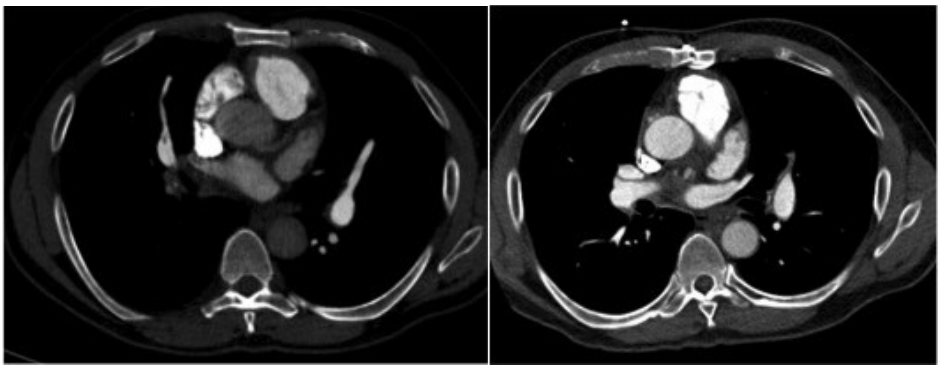

Figure 2: open superior segmental artery of the lingula pre-PEA (left image), occluded superior segmental artery of the lingula post-PEA (right image).

ICC17-77. 1-YEAR OUTCOME AFTER PULMONARY ENDARTERECTOMY FOR CHRONIC THROMBOEMBOLIC DISEASE

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Objective: Chronic Thromboembolic Disease (CTED) is a rare condition characterized by chronic thromboembolic lesions in the pulmonary arteries without pulmonary hypertension. It can causes dyspnoea and severe functional limitation. We describe a series of 30 patients with CTED treated with pulmonary endarterectomy (PEA).

Methods: From May 2008 to September 2015 we performed 30 PEAs for CTED. 13 (43%) were female. Mean age was 56 years (range 18 – 76). All patients were symptomatic for exertional dyspnea: 19 (64%) were in functional class WHO II, 7 (23%) in class WHO III, 4 (13%) in class WHO IV. Indications for PEA were: (1) presence of symptoms, (2) exercise O2-desaturation and (3) thromboembolic lesions occupying at least 40% of pulmonary vascular bed. Pre-operative and 1-year follow-up data were collected.

Results: 11 (37%) patients underwent monolateral PEA because of exclusively or predominantly unilateral lesions. 1 (3%) patient died for sepsis preoperatively and 1 patient died for respiratory failure 5 months after PEA. Overall survival was 93%. 1-year follow-up showed a marked improvement of WHO functional class: 22 (79%) patients were in class WHO I, 6 (21%) in class WHO II.

Hemodynamic and functional data are summarized in table.

	Pre-operative	1-year follow-up	p
6-minute walking test (m)	425 ± 105	474 ± 87	0.04
Modified Bruce Test (m)	350 ± 276	577 ± 312	0.007
Arterial pO2 (mmHg)	79.9 ± 11.2	84.4 ±10.8	0.08
Mean pulmonary pressure (mmHg)	19.8 ± 3.5	18.1 ± 5.3	0.24
Pulmonary vascular resistance (dyne*sec*cm-5)	244 ± 85	163 ± 66	0.001

Conclusions; PEA for CTED is a safe and effective procedure if performed in selected patients and in expert centers.

ICC17-78. IMPACT OF INHALATION OF NITROGEN OXIDE ON THE SURGICAL OUTCOME OF CTEPH

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Objectives: In PEA for CTEPH, there is a potential risk of residual PH causing both hemodynamic and respiratory deterioration in the perioperative periods. To prevent such adverse events and to improve the circulatory and respiratory conditions during and after PEA, inhalation of nitrogen oxide (NO) has been used. Its impact is investigated.

Patients and Methods: Between 2012 and 2016, 52 patients underwent PEA for CTEPH. For the recent consecutive 14 patients (INO group), NO inhalation was used at 20 ppm after the onset of the rewarming time after PEA, which was continued until the extubation in the ICU. The outcome was compared with that of the other 38 patients (non-INO group) of the earlier series without NO inhalation.

Results: In the non-INO group, 4 patients could not be weaned from CPB and required ECMO and IABP due to residual PH, one of who died from residual PH and pulmonary bleeding. Contrarily, all of the patients survived without ECMO support in the INO group. In comparison, there were some favorable tendencies of the hemodynamic parameters just after CPB weaning in the INO group; mPAP 26.4 (INO) : 27.2 (non-INO) mmHg, CO 4.5 : 4.1 L/min, PVR 321 : 340 dyne・sec・cm^{-5}. Extubation was also significantly earlier in the INO group: 20.1 : 46.8 hours.

Conclusion: It was demonstrated that NO inhalation has favorable impact on the surgical outcome of CTEPH due to relief of PH and related respiratory impairment after PEA.

ICC17-79. OUTCOME AFTER PULMONARY ENDARTERECTOMY FOR CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: RECENT 10- YEARS EXPERIENCE IN A SINGLE CENTER

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Objectives: Pulmonary endarterectomy for chronic thromboembolic pulmonary hypertension (CTEPH) has been a well-established procedure. This study reviewed mid-term outcome after pulmonary endarterectomy with recent 10- years experience.

Methods: Between 2007 and 2016, 100 patients underwent pulmonary endarterectomy for CTEPH. Mean preoperative pulmonary artery pressure and pulmonary vascular resistance were 46±10 mmHg and 1104±406 dynes/s/cm^{-5}. Mean follow-up period after surgery was 44 months (range, 1 to 115 months).

Results: In-hospital mortality was 3.0 %. In the 97 survivors, mean pulmonary artery pressure and pulmonary vascular resistance were significantly decreased to 24±11 mmHg and 429±313 dynes/s/cm^{-5} (p<0.0001, each case). Of 97 patients, 22 (22.7%) underwent balloon pulmonary angioplasty (BPA) for distal or recurrent thromboembolic disease. The mean duration between pulmonary endarterectomy and the initial BPA was 32.5±27.8 months. BPA showed significant decrease of mean pulmonary artery pressure (p=0.03) and pulmonary vascular resistance (p=0.02). The actual survival rate was 100% at 1 year and 98.5% at 5 years. There was no death related to CTEPH.

Conclusion: Pulmonary endarterectomy for CTEPH shows low mortality, and results in significant hemodynamic improvement. Combination therapy with balloon pulmonary angioplasty would contribute to favorable outcome for patients with distal or recurrent thromboembolic lesions.

ICC17-80. NEUROPSYCHOLOGICAL EFFECTS OF SHORT PERIODS OF SYSTEMIC CIRCULATORY ARREST IN PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION UNDERGOING ENDARTERECTOMY

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Objective: In our study we investigate whether the use of short periods of circulatory arrests during pulmonary endarterectomy (PEA) in patients with chronic thromboembolic pulmonary hypertension (CTEPH) have significant neuropsychological effects.
Methods: Between 2014 and January 2017, 70 of 165 18-to-80 years-old patients with CTEPH and who underwent PEA were evaluated with a neuropsychological protocol before surgery and three months after PEA. Also they were investigated for anxiety, depression and quality of life (QoL). In our Center, PEA is performed with 7-10 minutes periods of 24º C circulatory arrest interrupted by 5-7 minutes periods of reperfusion depending of measured of brain saturation. The average of the number of circulatory arrest is 10 SD ± 3 and the average time of total circulatory arrest is 85 SD ± 27.
Results: Three months after PEA, learning decreased from 40.86 ± 8.93 to 37.01 ± 8.67 and delayed memory changed from 9.42 ± 3.09 to 8.07 ± 3.13. Anxiety decreased from 7.70 ± 4.34 to 5.95 ± 3.64, depression reduced from 6.11 ± 3.49 to 4.48 ± 3.37 and quality of life improves. The perception of psychological well-being increases from 35.80 ± 8.63 to 45.08 ± 9.75 and perception of physical changes rises from 47.58 ± 10.70 to 54.75 ± 8.93.
Conclusions: PEA with prolonged total circulatory arrest time is a technique that does not worsens cognitive performance. Most of the investigated cognitive functions do not change at 3 months. In addition, anxiety and depression decreased while QoL improves.

ICC17-81. POSTOPERATIVE LEFT VENTRICULAR FUNCTION IN DIFFERENT TYPES OF PULMONARY HYPERTENSION: A COMPARATIVE STUDY

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Objectives: Temporary left ventricular (LV) dysfunction after pulmonary endarterectomy (PEA) for chronic thromboembolic pulmonary hypertension (CTPEH) is well described. True LV-failure has only been described after bilateral lung transplantation (bLTx) in pulmonary arterial hypertension (PAH) patients. We sought to identify factors that contribute to this LV-failure and hypothesized that an atrial septostomy before bLTx could prevent this LV-failure.
Methods: From our database, all PAH patients that underwent bLTx (n=24) and all CTPEH patients that underwent PEA, with a minimal reduction of 800 dynes.s.cm^{-5} (n=27), were selected. Perioperative demographic and echocardiographic data were analyzed.

Results: Results are depicted in tables 1-3. Pulmonary hypertension was diagnosed at significant younger age and time between diagnosis and surgery was significantly longer in PAH patients. PAH patients had significant larger right ventricular (RV) dimensions, but a similar preoperative LV diastolic dysfunction. Surgery caused significant decreases in RV dimensions, to a lesser extent in CTEPH patients, and significant increases in LV dimensions. Atrial septostomy for PAH showed a tendency to increase LV dimensions and cardiac index. Two PAH patients developed postoperative LV-failure. Their mean age at diagnosis, time between diagnosis and surgery, E/A ratio and LV-mass before bLTx were 10.9 years, 4776 days, 2.3, and 68 grams, respectively.

Conclusions: In PAH, diastolic dysfunction before surgery is comparable to CTEPH. However, age at diagnosis is younger and LV preload deprivation lasts longer. This might explain the occasional development of systolic LV-failure after bLTx. Preoperative atrial septostomy might train the LV, by increasing its preload, and avoid postoperative LV-failure.

Table 1 Demographic data.

	CTEPH - PEA (27)	PAH - bLTx (24)	p
Female/Male	9/18	11/13	0.4018
Age at diagnosis (years)	56.8 ± 14.3	37.4 ± 14.7	$< 1.10^{-4}$
Age at surgery (years)	57.9 ± 14.3	43.0 ± 13.4	$< 1.10^{-3}$
Time between diagnosis and surgery (days)	387 ± 440	2068 ± 1662	$< 1.10^{-5}$

CTEPH = chronic thromboembolic pulmonary hypertension, PEA = pulmonary endarterectomy, PAH = pulmonary arterial hypertension, bLTx = bilateral lung transplantation

Table 2 Pre- and postsurgical echocardiographic data.

	CTEPH - PEA (27)		PAH - bLTx (24)	
	pre	post	pre	post
LVEDD (mm)	37 ± 6	43 ± 6*	37 ± 7	43 ± 6*
LVESD (mm)	23 ± 5	29 ± 6*	22 ± 6	30 ± 6*
LV diastolic area (cm²)	20 ± 7	24 ± 7*	19 ± 5	24 ± 5*
IVS thickness (mm)	11 ± 2	11 ± 2	10 ± 2	11 ± 2
LVPW thickness (mm)	10 ± 2	11 ± 2*	9 ± 2	10 ± 3*
LV mass (g)	120 ± 42	168 ± 56*	104 ± 39	159 ± 53*
LVEF (%)	64 ± 11	65 ± 10	71 ± 11	65 ± 11*
E velocity (m/s)	0.45 ± 0.14	0.64 ± 0.19*	0.67 ± 0.21	0.75 ± 0.23
A velocity (m/s)	0.63 ± 0.18	0.66 ± 0.19	0.72 ± 0.16	0.58 ± 0.18*
E/A	0.80 ± 0.41	1.07 ± 0.53*	0.92 ± 0.31	1.44 ± 0.87
SV (ml)	50 ± 15	65 ± 19*	61 ± 15	65 ± 25
CI (L/min/m²)	2.37 ± 0.79	3.00 ± 0.87*	3.08 ± 0.84	2.75 ± 0.93
RV diastolic area (cm²)	31 ± 5	25 ± 5*	37 ± 10	18 ± 5*

LVEDD = left ventricular end diastolic diameter; LVESD = left ventricular end systolic diameter; LV = left ventricle; IVS = interventricular septum; LVPW = left ventricular posterior wall; LVEF = left ventricular ejection fraction; E = mitral peak early diastolic velocity; A = mitral peak late diastolic velocity; SV = stroke volume; CI = cardiac index; RV = right ventricle; * = significant difference with preoperative value; __ = significant difference with corresponding echo in PEA group.

Table 3 Echocardiographic data comparing PAH without and with AS.

	PAH-bLTx without AS (16)		PAH-bLTx with AS (6)		
	pre-bLTx	post-bLTx	pre-AS	pre-bLTx	post-bLTx
LVEDD (mm)	36 ± 5	43 ± 5[1]	32 ± 8	39 ± 5[3]	43 ± 5[3]
LVESD (mm)	21 ± 4	28 ± 5[1]	19 ± 5	24 ± 8	28 ± 4[3]
LV diastolic area (cm²)	21 ± 4	24 ± 6	14 ± 2[2]	19 ± 6	24 ± 6[3]
IVS thickness (mm)	10 ± 2	10 ± 2	12 ± 3	10 ± 2	12 ± 2[1]
LVPW thickness (mm)	9 ± 2	11 ± 3	9 ± 1	9 ± 2	10 ± 1
LV mass (g)	101 ± 30	156 ± 51[1]	144 ± 84	127 ± 52	181 ± 54
LVEF (%)	71 ± 12	67 ± 8	66 ± 4	71 ± 10	66 ± 5
E velocity (m/s)	0.64 ± 0.22	0.69 ± 0.19	0.54 ± 0.31	0.74 ± 0.23	0.84 ± 0.23
A velocity (m/s)	0.68 ± 0.17	0.58 ± 0.13	0.74 ± 0.21	0.85 ± 0.22	0.63 ± 0.36
E/A	0.95 ± 0.35	1.15 ± 0.35	0.73 ± 0.12	0.87 ± 0.31	1.34 ± 0.49[3]
SV (ml)	65 ± 18	66 ± 31	43 ± 8[2]	57 ± 9[3]	63 ± 17[3]
CI (L/min/m²)	3.25 ± 0.99	2.99 ± 1.10	1.98 ± 0.30[2]	3.07 ± 0.48[3]	2.51 ± 0.43[3]
RV diastolic area (cm²)	37 ± 10	18 ± 5[1]	41 ± 11	37 ± 8	18 ± 4[1,3]

PAH = pulmonary arterial hypertension; bLTx = bilateral lung transplantation; AS = atrial septostomy; LVEDD = left ventricular end diastolic diameter; LVESD = left ventricular end systolic diameter; LV = left ventricle; IVS = interventricular septum; LVPW = left ventricular posterior wall; LVEF = left ventricular ejection fraction; E = mitral peak early diastolic velocity; A = mitral peak late diastolic velocity; SV = stroke volume; CI = cardiac index; RV = right ventricle; [1] = significant difference with preoperative value; [2] = significant difference with pre-bLTx without AS condition; [3] = significant difference with pre-AS condition; no signifcant differences were found between PAH-bLTx without AS and PAH-bLTx with AS at the same time-points.

ICC17-82. SHORT AND LONG-TERM EFFECTS OF PULMONARY ENDARTERECTOMY ON VENTILATORY RESPONSES AND EXERCISE CAPACITY OF PATIENTS WITH CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION

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Rationale: Residual pulmonary hypertension (rPH) may affect long-term exercise improvement after pulmonary endarterectomy (PEA).

Objective: To evaluate the influence of rPH on short and long-term effects of PEA in patients with CTEPH.

Methods: 40 patients with CTEPH (40% male, 45±14yrs; 70% NYHA FC III/IV, cardiac index 2.1 ± 0.6L/min/m^2; pulmonary vascular resistance 1031±444 dynes.s.cm^{-3}) performed incremental cardiopulmonary exercise test before PEA and 30 pts after 6 and 12 months of procedure. rPH was evaluated by echocardiogram.

Results: 14 patients presented rPH and 6 of them died after surgery. The best CPET parameter to indicate rPH was a $\Delta VE/\Delta VCO_2 > 65$ [OR 8.3 (1.8-37)]. Clinical improvement was achieved in the first 6 months, even in patients with rPH (100% of patients in FC I/II). Peak VO_2 increased from baseline up to 6m (12±3 vs 15±4 mL.Kg.min, p 0.002) with a further improvement after 12m of PEA (18± 5mL.Kg.min, $p < 0.005$). In contrast, $\Delta VE/\Delta VCO_2$ decreased after 6m (65±18 vs 39±13, $p < 0.005$) without significant changes on 12m evaluation (37±11, p 0.32). Despite significant improvement, patients with rPH presented greater $\Delta VE/\Delta VCO_2$ than patients without rPH (80 ±14 at baseline; 56±17 at 6m, 54±11 at 12m).

Conclusions: Patients with CTEPH present a rapid reduction of excessive exercise ventilation after PEA, remaining stable thereafter, but exercise capacity continue to improve up to 12 months. Residual PH may influence long-term recovery.