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Combined pulmonary endarterectomy and balloon pulmonary angioplasty in patients with chronic thromboembolic pulmonary hypertension

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BACKGROUND: Pulmonary endarterectomy (PEA) is a curative treatment option for more than 60% of patients with chronic thromboembolic pulmonary hypertension (CTEPH). For selected inoperable patients, interventional balloon pulmonary angioplasty (BPA) has recently been established in addition to medical treatment. This approach disrupts scar tissue occluding the pulmonary arteries, leading to an improvement in parenchymal perfusion. CTEPH is occasionally heterogeneous, with operable disease on one side but peripheral, inoperable changes on the contralateral side. Performing unilateral PEA (on the operable side only) in these patients may lead to a worse hemodynamic outcome and increased mortality compared with patients who that can be surgically corrected bilaterally. We sought to determine the feasibility, safety, and benefits of BPA applied to the contralateral lung in several patients with predominantly unilateral disease that was amenable to treatment by PEA.
METHODS: Standard unilateral PEA in deep hypothermic circulatory arrest was performed in 3 CTEPH patients with poor pulmonary hemodynamics, and inoperability of the contralateral pulmonary artery obstructions was confirmed. The inoperable side was treated by BPA. The intervention was performed during the rewarming phase of cardiopulmonary bypass.
RESULTS: A dramatic improvement in pulmonary hemodynamics, with a mean reduction in pulmonary vascular resistance of 842 dyne · sec/cm^5, was achieved in all patients. World Health Organization Functional Class was also significantly improved at the midterm follow-up.
CONCLUSIONS: The combination of surgical PEA and interventional BPA is a new treatment option for highly selected high-risk CTEPH patients. A multidisciplinary CTEPH expert team is a basic pre-requisite for this complex concept.

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Chronic thromboembolic pulmonary hypertension (CTEPH) is a common form of pulmonary hypertension.[1] Up to 4% of patients who survive an episode of acute pulmonary embolism will develop CTEPH.[2,3] Proximal and

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distal stenoses and occlusions of pulmonary artery (PA) branches lead to PH with consecutive deterioration of right ventricular (RV) function and right heart failure. Increased PA pressure (PAP) and hyperperfusion of the patent vessels cause a secondary microvasculopathy and further clinical deterioration.[4] This vicious circle compromises the pulmonary and systemic circulation and is associated with a poor prognosis.

Patients with CTEPH should be examined and treated in a high-volume center by a specialized team of experts.[1,5–9] The gold standard treatment for CTEPH is pulmonary endarterectomy (PEA). This complex surgical intervention is often curative, with post-operative normalization of the pulmonary hemodynamics, and can be accomplished with a low risk for the patients in experienced centers.[5,6] However, up to 37% of the patients are deemed inoperable.[7] In many countries, medical treatment with the soluble guanylate cyclase stimulator riociguat has been approved for patients with inoperable CTEPH.[10,11]

In 2001, Feinstein et al[12] reported balloon pulmonary angioplasty (BPA) as a new treatment option for patients with inoperable CTEPH.[12] Recently, the procedure has been refined, mostly by Japanese centers, with improved clinical and hemodynamic results with a mean reduction of mean PAP (mPAP) from 43 mm Hg to 25 mm Hg.[13–17]

BPA is performed as a staged procedure to reduce the risk of reperfusion edema and to minimize the amount of injected contrast medium, depending on the patient's actual pulmonary hemodynamics and the number of PA lesions. However, BPA is not a competitive treatment option for operable CTEPH patients but seems to be a promising therapeutical tool for inoperable patients with subsegmentally located net-like structures or strands ("webs and slits") obstructing PA branches.[15,16]

There are rare cases, however, of technically operable obstructions on one side (mostly right-sided) that could be treated surgically combined with distal contralateral lesions not amenable to surgery that could be target areas for BPA. Depending on the severity of PH, low-risk patients may undergo PEA, and if needed, BPA during follow-up. For patients with very poor pulmonary hemodynamics, however, a staged procedure might carry an extreme perioperative risk of right heart complications and death. Therefore, PEA in combination with BPA of the inoperable side as a hybrid procedure might be a new therapeutic option for a carefully selected group of high-risk patients to instantly achieve a maximum reduction in RV afterload and to decrease the risk of RV failure after weaning from extracorporeal circulation. We report here 3 patients with severe CTEPH who were treated by combined PEA and BPA.

Pre-operative assessment

All patients were assessed by an experienced multidisciplinary team in an international CTEPH reference center. Clinical history, physical examination, 12-lead electrocardiogram, laboratory tests, echocardiography, cardiopulmonary exercise test, 6-minute walk distance, coronary angiography, right-sided heart catheterization, ventilation and perfusion scintigraphy, computed tomography (CT) angiography, and pulmonary angiography were performed. The results were assessed for all patients.

The 3 patients had bilateral distal PA obstructions that were considered technically operable on the right side with a high likelihood of technical inoperability of the subsegmental arteries of the left lower lobe. PEA was planned for all 3 patients with the possibility of combined BPA if surgical inaccessibility of the left-sided distal obstructions was confirmed.

Patient 1

A 68-year-old man with inoperable CTEPH was diagnosed in 2011, and combined PH-specific medication was initiated. After further clinical deterioration (World Health Organization Functional Class [WHO FC] IV), the patient was referred to our center for a second opinion. Pulmonary angiography showed lesions in almost every segmental branch of the right PA. On the left side, there were only a few subsegmental obstructions in the lower lobe. Right-sided heart catheterization revealed severe CTEPH (mPAP, 65 mm Hg; pulmonary vascular resistance [PVR], 1,600 dyne • sec/cm^5). The procedure was planned as an endarterectomy of the right PA branches and BPA of the left posterobasal segment if these lesions were deemed inoperable.

Patient 2

A 70-year-old woman with severe progressive dyspnea for 5 years was diagnosed with CTEPH in 2009. She denied further assessment of operability at that time, and PH-specific medication was initiated. Owing to further worsening of her clinical condition (WHO FC III), she was referred to our center. As in Patient 1, pulmonary angiography showed surgically accessible lesions in the right PA and sub-segmentally located obstructions in the left lower lobe with poor pulmonary hemodynamics (mPAP, 65 mm Hg; PVR, 1,630 dyne · sec/cm^5). A complete endarterectomy of 7 segmental arteries of the right side and BPA of the anterobasal and posterobasal segments (8 and 10) of the left PA was planned.

Patient 3

A 58-year-old man (WHO FC IV) was diagnosed with severe CTEPH (mPAP, 64 mm Hg; PVR 852 dyne · sec/cm^5). Comorbidity was significant, with a history of stroke and coronary artery disease. Pulmonary angiography showed exclusively segmental obstructions of the right PA with subsegmental lesions located in the left lower lobe and lingular PA branches (Figure 1). Endarterectomy of the right side for complete disobliteration with BPA of 3 segments of the left lower lobe (segment 8–10) and 1 lingular segment (segment 5) was planned. Coronary intervention or bypass was not indicated according to findings of the pre-operative coronary angiography.