Chronic thromboembolic pulmonary hypertension (CTEPH) is one of the leading causes of severe pulmonary hypertension. Although the diagnosis can be easily suspected by echocardiography and lung perfusion scanning, CTEPH is an under-recognized and frequently overlooked condition with a poor prognosis. Pulmonary endarterectomy (PEA) is an effective surgical procedure providing an acute and permanent relief of thromboembolic pulmonary hypertension and potential cure for most of the patients.

Based on high quality pulmonary angiograms, operability and indication for surgery have to be evaluated by an interdisciplinary team including an experienced surgeon for each individual patient. Pulmonary endarterectomy is a complex procedure: It resembles a true endarterectomy (not embolectomy) of the pulmonary artery branches down to the segmental and subsegmental levels. Extracorporeal circulation and periods of hypothermic circulatory arrest are mandatory. In specialized centres, the operative risk has been decreased to acceptable levels (< 10%). Long-term survival and quality of life are excellent after successful surgery even in patients with end-stage thromboembolic pulmonary hypertension and right heart failure.

Although CTEPH is primarily considered a major-vessel disease, secondary microvasculopathy develops in unaffected pulmonary artery branches over time. Therefore, earlier diagnosis and referral for surgery might further improve early and late results by reducing the severity of small vessel disease. As the procedure is complex, patients should be treated by a specialized multidisciplinary team.

**LITERATURE**