Pulmonary arteriovenous malformations (PAVM) should be treated by embolization

Pulmonary arteriovenous malformations (PAVM) are congenital vascular communications in the lungs. They act as shunts so that the blood running through these malformations is not oxygenated or filtered (Fig. 1). These patients will typically have dyspnea on exertion with lowered functional level, cyanosis and drumstick fingers, and are at high risk of emboli to the brain and other organs. Severe neurological events such as transitory cerebral ischemia, stroke, and cerebral abscess occur in 30%-40% of patients with untreated PAVM. There is also a risk of PAVM rupture, especially during pregnancy, with resulting severe bleeding. In more than 75% of cases, PAVM appear in patients with hereditary hemorrhagic telangiectasia (HHT) (Mb. Osler), and about 25%-30% of all patients with HHT have PAVM. The prevalence of HHT is estimated to be 1 per 5000-10 000, and the prevalence of PAVM in the population is about 1 per 20 000.

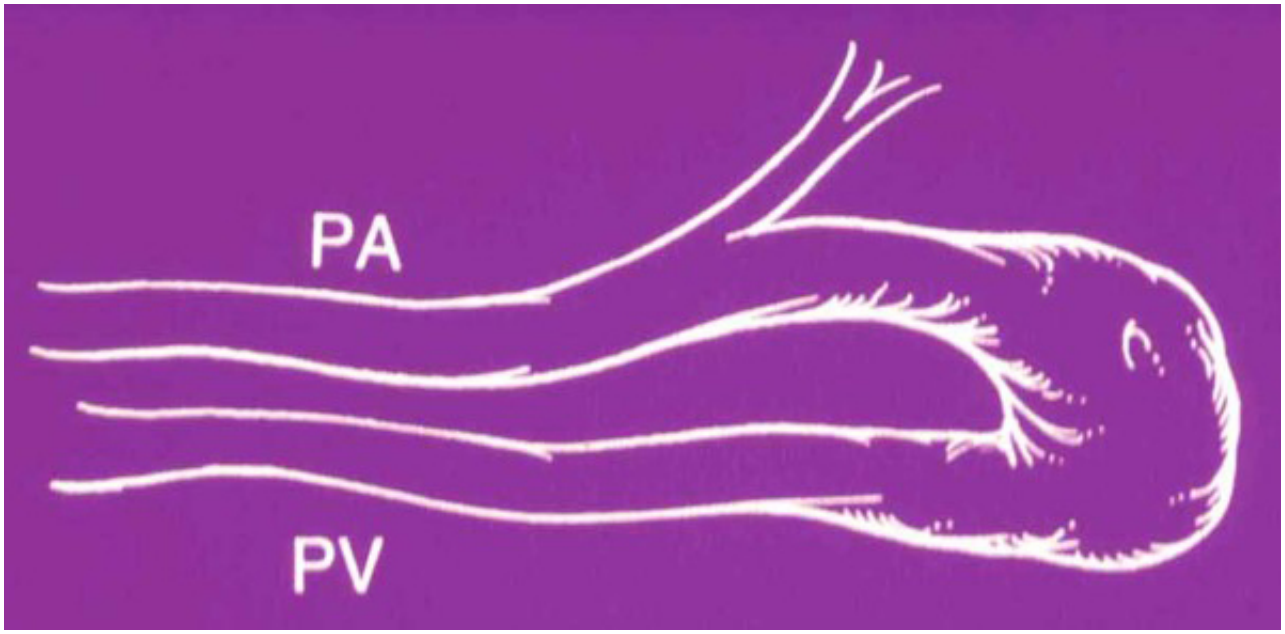

Fig. 1. Pulmonary arteriovenous malformations with shunting of the blood through the sacs without oxygenating or filtering the blood. PA = pulmonary artery. PV = pulmonary vein

The treatment strategy of first choice is embolization of the arteries supplying the PAVM. It is a minimally invasive procedure performed under local analgesia without need of convalescence, with high effectiveness and with few complications. The advantage of embolization is that the feeding artery to the PAVM is occluded and at the same time spares the adjacent normal pulmonary arteries (Fig. 2A and B). Embolization prevents cerebral stroke and abscess as well as pulmonary hemorrhage and further raises the functional level and thus there is evidence-based indication to

treat these malformations. Technical success with occlusion of the feeding artery is achieved in about 95%-100% of cases in experienced hands. Re-canalization or primary insufficient embolization has, however, been described in about 8%-15% of cases. Adverse effects of this therapy are not severe and are seen in about 10%-20% of cases. Self-limited pleurisy with or without a small pulmonary infiltration and respiratory chest pain is seen in 10%-15%, precordial pain in 2%-5%, primary coil dislocation or malpositioning during delivery in 4%. Catheter-induced bradycardia and ectopic heart beats are common during catheterization but are self-limiting. Mortality has never been reported in relation to this treatment. Clinical and anatomical evaluation after embolization of PAVM is important to detect persistent or re-perfused lesions. These patients will often have symptoms but a significant minority are asymptomatic.

The aim of the study was to assess the clinical outcome of patients after embolization of PAVM. The study included all patients with PAVM treated with embolization at a reference center for HHT and PAVM undertaken over a 20-year period. Demographic data, HHT genotyping, clinical presentation, and outcome were registered. Patients with HHT were compared to the patients without HHT. Clinical examination, contrast echocardiography, and computed tomography (CT) were used to assess the clinical outcome at follow-up. One hundred and thirty-six patients with 339 PAVM underwent embolization during the study period: 22 did not have HHT. Solitary PAVM were more common among patients without HHT than with HHT. Mean follow-up after the first embolization was 58 months.

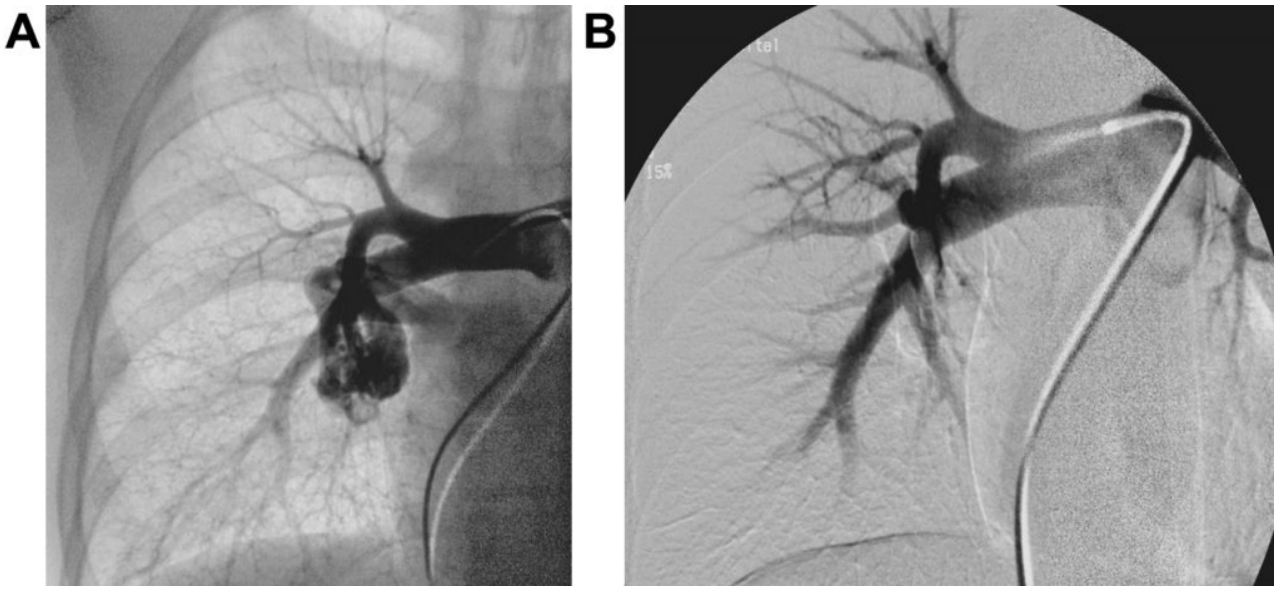

Fig. 2. Big PAVM before (left picture) and after (right picture) embolization with occlusion of the feeding artery to the sac. Normal vessels are spared.

Mean age at first embolization was 46.5 years, and at last follow-up 51.8 years. The clinical

success without shunt at follow-up was 87%. The 30-day mortality related to the embolization was 0%. Most patients could be treated during one session, but many will need a long follow-up with repeated clinical examinations and re-embolization. Most of the patients referred for embolization of PAVM had HHT. Multiple PAVM is associated with HHT. Patients with PAVM should be screened for HHT and patients with HHT for PAVM. Embolization is a safe procedure with high clinical success. Embolization of PAVM is a definitive treatment and is a well-established method with a significant effect on oxygenation of the blood and prophylactic effect on paradoxical emboli to the brain. Screening for PAVM in patients at risk is recommended, especially in patients with HHT.

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Publication

Pulmonary arteriovenous malformations: a radiological and clinical investigation of 136 patients with long-term follow-up.
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