**Article\***

"Development of the European Network in Orphan Cardiovascular Diseases"

„Rozszerzenie Europejskiej Sieci Współpracy ds Sierocych Chorób Kardiologicznych”

 **Title: Treatment of pulmonary artery aneurysm**

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**Date:29.09.2014**

The optimal treatment of pulmonary artery aneurysm has not been established.

In patients with normal pulmonary artery pressure the risk of rupture is low therefore conservative treatment seem reasonable. In patients with pulmonary arterial hypertension treatment should include diuretics, anticoagulants, and pulmonary vasodilators such as endothelin receptor antagonists, phosphodiesterase type 5 inhibitors, and prostacyclin derivatives. Nevertheless the risk of dissection and rupture in patients with pulmonary hypertension is high. On the other hand they have also a high surgical risk. When pulmonary artery aneurysm results from vasculitis it can be treated with immunosuppressive drugs and the effects of the treatment should be strictly monitored

Interventional treatment includes coil embolization of aneurysms of small branches of pulmonary artery, there are also some reports of occlusion of a dissected pulmonary artery aneurysm by a covered stent. Patients with aneurysm of central pulmonary arteries will require surgery. Indications for surgery include: absolute pulmonary artery aneurysm diameter ≥5.5 cm, increase in the diameter of the aneurysm of ≥0.5 cm in 6 months, compression of adjacent structures, thrombus formation in the aneurysm sack, appearance of clinical symptoms, evidence of valvular pathologies or shunt flow, signs of rupture or dissection.

Aneurysmectomy and repair or replacement of the right ventricular outflow tract is considered as a method of choice The most common procedure is the replacement of the PA and the pulmonary trunkwith a conduit starting in the right ventricular outflow tract.

Perioperative morbidity is comparable to that of the repair of

aneurysms of the ascending aorta.

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