Pulmonary artery aneurysm in pulmonary arterial hypertension associated with congenital heart disease

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Introduction: Pulmonary artery aneurysms are a common finding in patients with congenital heart disease, often asymptomatic and with a described low complication rate. The aim of this study is to assess the prevalence of PAA in a population with pulmonary arterial hypertension associated with congenital heart disease (PAH-CHD) and associated complications.

Methods: All adult patients with PAH-CHD at our institution with angiography of the pulmonary tree by computed tomography (CT) or magnetic resonance imaging (MRI) were included in a retrospective analysis. PAA was defined as a main PA diameter > 40 mm.

Results: In this study, the frequency of PAA was as high as 76%. Simple pre-tricuspid lesions had larger diameters than post-tricuspid lesions (p=0.004), but there were no significant differences with complex lesions. Also, no differences were found between the median diameters of the different PAH-CHD groups. No relationship was seen between the development of aneurysms or larger diameters and haemodynamic severity, even a majority of patients had severe pulmonary arterial hypertension.

15 patients (25% of total with PAA) had complications related to PAA: 4 patients with PA dissection (isolated or with associated thrombosis), 7 patients with chronic pulmonary thrombosis and 5 patients with left main coronary artery compression (figure 2). There were no complications in patients with diameters less than 40 mm. During a median follow-up of 3.8 years (IQR 1.6 to 6.4 years) 24 patients (30%) died or needed lung/cardipulmonary transplantation (17 and 7 respectively). Only two (8.3%) of the patients with final events did not have PAA. Survival analysis (figure 1) showed a higher rate of death or need for transplantation in patients with pulmonary artery aneurysm, especially if there was an added complication (log-rank test, P=0.017).

Conclusions: Pulmonary artery aneurysms in patients with pulmonary arterial hypertension associated with congenital heart disease are common. The rate of related complications is also high in this population and may have prognostic implications.
Complications related to Pulmonary Artery Aneurysm

- Isolated PA dissection: 5
- Isolated PA thrombosis: 2
- PA dissection and thrombosis: 5
- Left main coronary artery compression: 2
- Hoarseness secondary to recurrent laryngeal nerve palsy due to compression: 1

Complications related to PA