

## MP5-8

### Outcomes of peripheral pulmonary artery branch stenoses

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#### Background

Peripheral pulmonary artery branch stenosis (PPAS) is defined by the narrowing of pulmonary arteries after the hilum. Outcomes are scarcely known and therapeutic alternatives are limited.

#### Objectives

We sought to describe the different types of PPAS, and their respective outcomes.

#### Methods

Over a period of 10 years, all patients with PPAS were included into the study. Clinical, ultrasound and hemodynamic data were collected at the time of diagnosis. Only patients with biventricular repair of CHD were included. Patients were classified into 3 groups (see figure): group 1 with diffuse hypoplasia from segment 1 to segment 4 or 5; group 2 with PPAS of segment 3 branches with or without aneurysms; and group 3 PPAS of segment 4 and 5.

#### Results:

45 patients were included: 11 group 1, 31 group 2, 3 group 3. One third had associated congenital heart disease. At time of diagnosis, 50% were under 5 years, and 80% were asymptomatic. Mean right ventricle/left ventricle (RV/LV) pressure was 0.99 and was not different between groups. 7 patients had percutaneous angioplasty and none was treated surgically. Median follow-up was 6.3 years. No changes in RV/LV pressure ratio was observed during follow-up. Survival at 10 and 20 years was 95.2% and 66.7% with no difference between groups. The presence of symptoms and RV pressure > 110 mmHg at diagnosis were significantly associated with right heart failure, hemoptysis, syncope and pulmonary branch thrombosis ( $p=0.003$ ;  $p=0.04$  respectively). Right heart failure and dyspnea on exertion at diagnosis were associated with death ( $p=0.01$  for both).

#### Conclusion:

PPAS is a rare and severe disease since one third of patients die in childhood. The anatomy of pulmonary branches did not predict outcome. The presence of symptoms and suprasystemic RV pressure are predictors of outcome.

