Plastic Bronchitis - Symptomatic improvement after pulmonary arterial stenting in four patients with Fontan circulation

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Introduction: Plastic bronchitis (PB) is a severe complication after a Fontan procedure with an estimated incidence around 1-2%. If not treated adequately the prognosis of PB in cardiac patients is very poor. We present four patients with PB after a total cavopulmonary connection with a stenosis of the left pulmonary artery that was stented successfully. In three patients symptoms of PB improved after the catheter intervention.

Methods: Four patients (cardiac diagnoses: hypoplastic left heart syndrome 2, tricuspid atresia 1, complex cyanotic heart disease with a functionally univentricular heart 1) with PB at a median age of 31 month (range 21.5 – 45 month) with a median weight of 12.7Kg (range 9.5-14.8Kg) presented at our clinic.

A complete separation of the pulmonary and systemic circulation by an extracardiac non fenestrated total cavopulmonary connection had been performed at a median of 11 months (18-21 month) preceding the hospital admission. All patients underwent a complete cardiological examination including cardiac catheterisation.

Results: All four patients had significant LPA stenosis, which was treated by stent implantation. The LPA-diameter was augmented from a median value of 4.8 mm (range 3.9-6mm) to a median value of 8.7mm (range 7.8-10.4mm). The median transpulmonary gradient was 8 mmHg (range 6-9 mmHg) and the median central venous pressure was 15mmHg (range 12-17mmHg). Until today three children are free of symptoms. Solely one child, the one with the highest transpulmonary pressure, redeveloped casts. Median follow-up time is 13.5 months (range 8-25 month).

Conclusions: This series of cases shows significant improvement of PB after successful LPA-stenting in at least 3 patients. Our cases show that haemodynamic improvement is crucial in the management of PB. Any effort should be undertaken to relief any pulmonary artery obstruction in these patients before fenestration of the Fontan tunnel or cardiac transplantation are performed alternatively.