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Plastic bronchitis after extracardiac total cavopulmonary connection

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Introduction

Plastic bronchitis (PB) is a severe complication in patients with Fontan circulation. The incidence of PB is low and only few reports are available. The aim of our study was to investigate the perioperative factors at the time of total cavopulmonary connection (TCPC), which might prove predictive for the later onset of PB; as well as the clinical status and the medical course of the patients with PB.

Method

A retrospective cohort study included all patients (Jan 2000 – Dec 2012) who received TCPC, using an extracardiac conduit for a functionally univentricular heart at the German Heart Centre Munich. Patients who developed PB during follow-up were included in "PB group" while the remaining patients constituted the "non-PB" group.

Results

The prevalence of PB at our institution was 2,4% (8/333). The median time interval between the TCPC and the first diagnosis of PB was 29 months (range 2 weeks - 113 months). At the time of TCPC, there was no significant difference in pre-operative weight, age, central venous pressure, transpulmonary gradient and arterial - or venous oxygen saturation between the two groups. The occurrence of post-operative chylothorax was significantly higher in the PB group (7/8 in the PB group vs. 64/325; $p < 0.001$), as well as the occurrence of diaphragm paresis (4/8 in the PB group vs. 25/325; $p < 0.001$). On hospital admission for symptomatic PB 6/8 patients had an underlying problem leading to impaired hemodynamics: severe aortic valve regurgitation in two patients, severe left pulmonary artery (LPA) -stenosis in four patients. One patient developed cardiac decompensation after pneumonia and in one patient no detectable cause for PB was found. One patient received an aortic valve replacement, four were treated interventionally by LPA-stenting, while the remaining patients received a conservative treatment. During follow-up (median time 18 months (range 5-32 months), six patients are free of PB symptoms, one patient died and one patient is symptomatic with PB.

Conclusions

Post-operative chylothorax and diaphragm paresis at the time of TCPC were significantly more frequent in patients who later developed symptomatic PB. Compromised hemodynamics was found in nearly all symptomatic patients.