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Plastic bronchitis as a late complication in children after Fontan operation

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The etiology of plastic bronchitis (PB) as a late complication after Fontan operation is still unclear. Patients: In our center out of 280 Fontan patients (median follow-up 4,2 years (2 mo - 14,5 years) 7 children (5 girls, 2 boys) developed PB (2,5%). Six had HLHS and 1 a single ventricle with hypoplastic arch. All underwent 3-stage palliation with Norwood- (Sano: 5, BT-Shunt: 2), Glenn- (median: 3,6 months (2,5-5,3) and extra cardiac Fontan OP (median: 3,3 years (2,3 - 7,2)), 5 of them fenestrated. Results: Onset of PB happened at a median age of 7,3 years (3 - 12,4) or 1,1 years (0,2 - 10,1) after Fontan OP. Transient postoperative chylothorax was present in 5 patients. Therapy of PB consisted of bronchoscopy (n=7) ECMO (n=1) and cardiac interventions to improve hemodynamics (n=5) as well as inhaled rt-PA, steroids and medication to improve cardiac function, to reduce pulmonary vascular resistance. After a follow-up of median 3,1 years (1,4 - 8,5) all patients are alive. One child with heart failure and tricuspid regurgitation was transplanted and has no casts 4 months after HTX. 2 patients are cast free without rt-PA inhalations for 1,5 years following successful interventional treatment of hemodynamic problems with increased CVP (APCAs and AV fistula), 1 patient is now cast free for 2,5 years without therapy after re-opening of the fenestration and pacemaker implantation, 3 patients are still producing casts for 3, 3,1 and 10 years respectively. One 15 year old girl without lymphatic anomalies responding to high dose cortisone, one 7 year old girl with an obvious lymphatic fistula from the thoracic duct to the right lung (awaiting selective lymphatic intervention) and one 13 year old girl after 11 interventions including re-fenestration, repeated APCA coiling and isthmus-stenting now awaiting lymphatic imaging.
Conclusion: PB after Fontan remains a diagnostic and therapeutic challenge as some etiologies are still unknown and multifactorial and each patient has specific characteristics needing tailored treatment.