

Need of ECMO in a reversible life-threatening plastic bronchitis 8 years after Fontan completion

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Introduction: Plastic bronchitis is a known but rare complication in patients with failing Fontan circulation, because of high central venous pressure, elevated transpulmonary pressure or pump failure. Bronchial casts with rubber-like consistency develop in the tracheobronchial tree and cause airway obstruction. The patients develop wheezing and desaturation and in case of ventilator therapy they need high mean airway pressures for oxygenation and therapeutic bronchoscopy to remove the casts.

Case: We report of a 12 year old boy in good clinical condition 8 years after total cavopulmonary connection. In a routine echocardiography an enlargement of the ascending aorta and aortic valve regurgitation was noticed without clinical symptoms. After replacement of the aorta ascendens and aortic valvuloplasty he showed a hard postoperative course with life-threatening lung haemorrhage, what was treated by transfusion of fresh frozen plasma and recombinant factor seven. Some days later he developed severe plastic bronchitis of the left lung. The patient got too hypoxic and hemodynamically unstable under high mean airway pressure for therapeutic bronchoscopy, so we took him on extracorporeal membrane oxygenation (ECMO) and removed the bronchial casts in stable condition. After diagnosing high degree of aortic valve regurgitation in the cath lab, the aortic valve was replaced. Two days later after normal bronchoscopy we weaned the patient from ECMO successfully. One year after the operation he is still free of plastic bronchitis and again in good clinical condition.

Conclusion: Almost one third of the reported Fontan patients with plastic bronchitis died of respiratory failure within one year, the rest suffer on recurrent episodes of respiratory compromise under different therapy (e.g. sildenafil, inhalation of recombinant tissue plasminogen activator). In case of an acute postoperative problem in Fontan circulation the occurrence of plastic bronchitis seems to be reversible, so each therapeutic approach, ECMO included, is warranted.