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Around *PediHeart:* Plastic Bronchitis

Over the past year or two there have been a number of interesting cases submitted to the *PediHeart* bulletin board regarding a phenomena referred to in the literature and by *PediHeart* members as "plastic bronchitis". I felt that this would be a good time for *Around PediHeart* to summarize those postings. When associated with congenital heart disease "plastic bronchitis" is characterized by acellular fibromucinous bronchial casts that cause symptomatic airway obstruction. Similar casts are also seen conditions that cause inflammation of the bronchopulmonary tree but these casts tend to be made up of densely packed inflammatory (eosinophils) cells [1].

There were 8 cases presented to the group. No cardiologist had more than one case. All instances of plastic bronchitis occurred after a lateral tunnel type Fontan procedure with fenestration. For most patients the onset of symptoms was roughly 2--3 years after the Fontan. For one patient it began 6 months post Fontan. Three members included hemodynamic data. In one, the mean Fontan pressure was 13 mmHg with a wedge pressure of 10 mmHg and a left ventricular end diastolic pressure (LVEDP) of 5 mmHg. This patient was 6 months post Fontan and no therapy for the plastic bronchitis was successful including mucolytics, and Coumadin. The second case with available data was 18 months post-op and had a Fontan pressure of 13 mmHg and an LVEDP of 8 mmHg. Again no treatment was successful. The last case with data was several years post-op from his Fontan. He had a mean Fontan pressure of 16--18 mmHg and an LVEDP of 8 mmHg. In this instance resolution of plastic bronchitis occurred with aggressive medical treatment using continuous outpatient Milrinone infusion to improve ventricular function and lower the Fontan mean pressure. While hemodynamic data was not available for the other cases one had hypoalbuminemia and another had heart failure and protein losing enteropathy a year after presenting with plastic bronchitis. In another case the plastic bronchitis occurred twice after two subsequent non-cardiac surgeries years apart. Treatments temporally associated with improvement included Milrinone infusion, thoracic duct ligation, steroid administration, cardiac transplantation, heparinization, and doing nothing. Treatment failures included anticoagulation therapy with Coumadin, coil embolization of pulmonary collateral vessels, inhaled mucolytics and bronchodilators, and doing nothing.

Plastic bronchitis is a real entity and, given that eight cardiologists from different centers posted a case on *PediHeart*, it may not be so very rare. The mechanism for plastic bronchitis may involve an elevation in systemic venous or pulmonary venous pressure [2]. This was the presumed cause behind the patient who improved with Milrinone therapy and perhaps the patient who later developed protein losing enteropathy and heart failure. It may also involve a primary abnormality in lymphatic drainage as suggested in the literature [1] and by the patient who improved after thoracic duct ligation. Finally, given that in some patients plastic bronchitis occurred and spontaneously resolved without reason or intervention, it would be safe to say that, while this condition is interesting, the jury is still out.

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