Plastic Bronchitis Occurring Late after the Fontan Procedure in a Child: Treatment with Stent Implantation in the Left Pulmonary Artery

Ebru Yalcin, MD, Ugur Ozcelik, MD, Alpay Celiker, MD

Plastic bronchitis is characterized by marked obstruction of the large airways by bronchial casts. Bronchial casts take the shape of the bronchi of a lobe or a lung. These casts may result in mild symptoms or life-threatening disease, and diagnosis is usually made when casts are expectorated or removed by bronchoscopy. Casts are often a complication of underlying bronchial diseases associated with mucus hypersecretion, such as cystic fibrosis, asthma, allergic bronchopulmonary aspergillosis, bronchiectasis or bronchopulmonary infections. Congenital and acquired cardiopathies have also been implicated in the formation of bronchial casts. Nevertheless, this disorder has also been shown to occur in children with congenital cyanotic heart disease who have undergone cardiothoracic surgical procedures, most commonly the Fontan procedure. In 1997, Seear, et al.3 separated bronchial casts into two well-defined groups as pathological: Type 1 (inflammatory) group, consisting of casts composed mainly of fibrin with a dense eosinophilic inflammatory infiltrate; and type 2 (acellular) group, consisting of casts composed mainly of mucin with little or no cellular infiltrate, which occurs mostly in children with congenital cyanotic heart disease with palliative surgery. Type 1 casts seem to be well controlled with bronchoscopic removal of the cast and steroid treatment, but treatment of type 2 casts has been more problematic: optimal treatment for patients with type 2 casts is not clear, unsuccessful, and the prognosis probably depends on the underlying cardiac status in this group.3

We report a child with tricuspid atresia who had undergone the Fontan procedure and developed plastic bronchitis 5 years after surgical repair. Some narrowing at the left pulmonary artery was detected, thus, stent implantation was performed in this region. Such long-term improvement in a patient with plastic bronchitis following stent implantation has not been previously reported.

Case Report. A girl with tricuspid atresia had undergone the Fontan procedure at 2.5 years of age. Neither pleural nor pericardial effusion developed after the post-operative period. The following 5 years were uneventful. At the age of 7.5 years, she complained of coughing spells that often resulted in the expectoration of bronchial casts (Figure 1). She had no history of allergies or asthma, and her chest X-ray was normal. Flexible bronchoscopy was performed, which revealed unremarkable findings. The casts consisted of mucin and fibrin with few mononuclear cells. She did not improve with acetylcysteine (p.o.) and chest physiotherapy over a 4-week treatment period. The patient continued to have more frequent episodes of cast expectoration. Echocardiography was unremarkable and abdominal ultrasound revealed a minimal increase in the calibre of the inferior vena cava and hepatic veins. Cardiac catheterization showed some narrowing due to a kink at the entrance of the left pulmonary artery (Figure 2), thus, stent implantation was planned. A Palmaz 18 balloon expandable stent was crimped on a 10 mm Tyshak balloon and implanted in the origin of the left pulmonary artery (Figure 3). Before implantation, the patient’s mean pulmonary artery pressure was 13 mmHg, the mean right pulmonary artery pressure was 13 mmHg, and the mean left pulmonary artery pressure was 10 mmHg. After stent implantation, the pressures were measured as 11 mmHg in the pulmonary artery and 10 mmHg in the left pulmonary artery. Before stent implantation, in perfusion scintigraphy, the left lung contributed only 34% to the total lung function and the right lung contributed 66%. Three days after stent implantation, significant improvement was not observed in these ratios. The child improved following stent application. The patient is doing well 16 months after this procedure, and continues to expectorate bronchial casts, but much less in terms of quantity and frequency.

Discussion. Plastic bronchitis has also been shown to occur in patients who have undergone cardiothoracic surgical procedures, most commonly the Fontan procedure. The Fontan procedure is performed on children with congenital single-ventricle heart disease and entails the diversion of systemic venous return into the pulmonary circulation. This operation has been associated with recurrent bronchial cast formation in 8 children reported in the last 15 years. The mechanism by which this procedure predisposes children to cast is not obvious. Langevin, et al. explained a physiopathological mechanism for the formation of bronchial casts. Endobronchial lymphatic leakage, surgical trauma to the lymphatic channels surrounding the bronchi, pleural adhesions, lymphatic dysplasia and high systemic venous blood pressure are the principal factors resulting in the formation of bronchial casts in patients with cardiopathy.

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changes occur as a result of increased central venous pressure, and surgical trauma to the lymphatic channels surrounding the bronchi may be responsible for the occurrence of casts in these cases.

Our patient did not have any evidence of pleural effusions in the post-operative period, and she developed plastic bronchitis 5 years after the surgical repair. Her cardiac functions were normal, and presence of systemic venous hypertension and hepatic congestion were revealed by abdominal ultrasound. The Fontan operation causes an increase in systemic venous pressure in some cases, which not only obstructs lymph drainage from the thoracic duct, but also causes hepatic congestion, which significantly increases the volume of lymph in the thoracic duct. Some authors speculate that the formation of bronchial casts in children who have undergone the Fontan procedure is another manifestation of the unique univentricular circulation that predisposes these patients to third-space fluid loss; in particular, effusions, ascites, and peripheral edema are characterized by systemic venous hypertension. Ventricular systolic or diastolic dysfunction, atrioventricular valve insufficiency, and a variety of arrhythmias and protein-losing enteropathy are all reported to predispose a patient to and potentiate these problems. Nevertheless, these findings were not detected in our patient.

Brogan, et al. reported a 5-year-old boy with tricuspid atresia who underwent a fenestrated Fontan procedure with an intracardiac tunnel. Post-operatively, he developed bilateral pleural effusion (which resorbed spontaneously), atrial node dysfunction, and pericardial effusion. Five months after the Fontan procedure, plastic bronchitis developed and 6 months later, cardiac catheterization showed good function and some irregularity at the origin of the right pulmonary artery. One year after his Fontan operation, the boy underwent repeat cardiac catheterization for placement of a pulmonary artery stent and an AV pacemaker, and he experienced no episodes of cast expectoration or hypoxemia. This patient might have benefited from the placement of a pulmonary artery stent and/or AV pacemaker, but the authors did not discuss this matter in their report.

McMahon, et al. reported a child who presented with recurrent expectoration of bronchial casts 6 months after a fenestrated lateral tunnel Fontan circulation for pulmonary atresia. Cardiac catheterization demonstrated elevated central venous pressure...
(CVP) with two areas of stenosis within the Fontan circuit, specifically at the junction of the right superior caval vein and the right pulmonary artery, and between the atrial baffle and the right superior caval vein. Insertion of Palmaz stents in these areas resulted in a reduction in CVP and a transient reduction in the production of casts. Eight weeks after catheterization, the patient developed respiratory arrest and died.

Multiple treatment modalities have been used in the management of plastic bronchitis. Different therapies have been applied in various reports, depending on the nature of cast.

Corticosteroids have been employed preferentially with type 1 casts, while bronchoscopic removal has been routinely practiced in most cases. Acetylcysteine, chest physiotherapy, systemic and inhaled corticosteroids, DNAse and aerosolized urokinase have all been used. Though the optimal treatment for patients with type 2 casts is not clear; reducing venous pressure and improving cardiac output in children with underlying cardiac defects are considered important and potentially useful.

In our patient, the high systemic venous pressure arose from both the Fontan operation and left pulmonary artery occlusion, which improved via stent implantation in the left pulmonary artery. We hope that our experience will provide some help in the management of children with type 2 bronchial casts.

We believe that, once identified, a careful hemodynamic evaluation should be made to identify treatable causes of systemic venous hypertension, rather than only directing efforts at lysis of casts in these cases.

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