Review of the Effectiveness of Tissue Plasminogen Activator for the Treatment of Plastic Bronchitis in Patients with Fontan Physiology

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Plastic bronchitis is a condition characterized by the formation of bronchial casts ranging from small to large obstructions of the pulmonary airways. Cast formation is spontaneous and resembles a plastic, rubbery model of the airways it obliterates. Its form was first described by Galen (A.D. 131-200). One of the patient populations at risk for developing this rare complication are those with single ventricle disease who are surgically corrected to Fontan physiology. It is a very abnormal type of circulation that is unique to the development of plastic bronchitis. The cellular composition of plastic bronchitis differs from the casts seen in patients with chronic respiratory diseases such as asthma or idiopathic pulmonary fibrosis. The casts are seen in an aerosolized form. Patients with primary pulmonary hypertension may develop plastic bronchitis. A review of patients with Fontan physiology in an aerosolized form has shown to be effective therapy in most cases. The following information is an update on the adaptation of this therapy in the last 5 years.

Pathophysiology

Plastic bronchitis, or cast formation, involves looking upon systemic ventricle return directly to the pulmonary artery (rare) bypassing the right side of the heart. This is also referred to as "passive circulation" and forward blood flow through the lungs is accomplished by pressure gradients between the central venous circulation and the pulmonary vascular bed. The endothelium is altered, however increased pulmonary venous pressures may contribute to affecting the respiratory epithelium and interruption of the bronchial mucus-carrying pancreatic material to leak and dysmucosification of mucus with resultant cast formation. Histological samples of patients with congenital heart disease reveal acellular casts with mucin and fibrin and the absence of Charcot-Leyden crystals. Patients with primary pulmonary disease reveal a combination of inflammatory casts, eosinophils, Charcot-Leyden crystals and mucus thought to be due to Influenza bronchial inflammation (Do et al., 2009; Goldberg, 2010; Singhal, 2011). See figure 1.

Signs and Symptoms

The most common presenting symptoms are cough and interstitial dyspnea (Goldberg, Rychik, & Dados, 2000). The cough may be productive or non-productive depending on the patient's ability to expectorate casts. Frequently, thick mucus is expectorated with a mucous plug consistency which can be decaying as the mucous plug may actually become detached from the bronchial wall and break away from the cast. Chest pain and fever are also reported but occur more frequently in patients with pulmonary parenchymal disease. Cardiac patients rarely exhibit a fever, and blood and sputum cultures are usually negative (Heath et al., 2011). Breath sounds may be diminished, coarse, and wheezing may be present. Chest x-ray reveals opacification and consolidated fields similar to pneumonia and bronchitis or expectoration of a cast is the only reliable determinant of the diagnosis. See figure 2.

Treatment

Once the airway is stabilized, patients with congenital heart disease suffering from the rare complication of plastic bronchitis are first evaluated for circulatory failure. A candard catheterization is performed to determine venous systemic pressures, development of collateral circulation in the lungs, narrow pulmonary vessels and evaluation of the Fontan circuit. The Fontan conduit may be fenestrated to reduce systemic venous pressure; narrowed blood vessels may be stented, open, collateral closed with obstructive ducts and the functioning cardiac valve and ventricle assessed for optimal performance. Fontan physiology, at its best outcome mimics chronic, low grade congohestial failure. Every effort is made to decrease systemic venous pressures and optimize cardiac output (Goldberg, et al., 2010). However, there is no guarantee of successful prevention of a recurrence of plastic bronchitis. If histology shows dilated lymphatics, thoracic duct ligation is also considered especially if the patient exhibits problems with pleural effusions. (Colaner, Quart, Pozzi, Gaughr, Crump, & Heath, 2010). Pharmacological therapy interventions include systemic and inhaled corticosteroids, mucolytics such as acetylcysteine and dextrose alcohols, and fibrinogens namely urokinase and Alteplase.

Tissue plasminogen activator (tPA) or Alteplase, is a serine protease that leads to localized fibrinolysis by converting plasminogen to plasmin. According to the literature topical application and nebulized tPA has been the most successful treatment without adverse effects in several case reports of children with congenital heart disease complicated by cast bronchitis. Bregen et al. (as cited by Do et al, 2012) looked at the acute phase of illness and typically applied urokinase. Alteplase and the saline provided no effect, the Urokinase made the cast softer and, the solution treated with Alteplase, completely dissolved. See figure 3.

The left picture shows cast fragments wedged in the upper and lower lobes. The picture on the right shows the effect of tPA application 15 minutes. Copyright 2012 American Academy of Pediatrics.

Conclusion


Additional Resources


Tissue plasminogen activator (tPA) or Alteplase, is a serine protease that leads to localized fibrinolysis by converting plasminogen to plasmin. According to the literature topical application and nebulized tPA has been the most successful treatment without adverse effects in several case reports of children with congenital heart disease complicated by cast bronchitis. Bregen et al. (as cited by Do et al, 2012) looked at the acute phase of illness and typically applied urokinase. Alteplase and the saline provided no effect, the Urokinase made the cast softer and, the solution treated with Alteplase, completely dissolved. See figure 3.

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