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 intricate and resembles a plastic, rubbery model of the airway it obstructs. 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 congenital heart disease who are surgically corrected for 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 in terms of how these casts develop and, as a result, complete airway obstruction may occur more quickly with a resultant life-threatening and possibly fatal event. Bronchial casts may also be seen in other disease states such as sickle cell disease, metastatic lung tumors, bronchopulmonary aspergillosis, tuberculosis, and alpha and shotgun injuries (Do, Randhawa, Chin, Parsapour, & Nussbaum, 2012). Tissue plasminogen activator in an aerosolized form has shown to be effective in most cases. The following information is an update on the adaptation of this therapy in the last 5 years.

Pathophysiology

Fontan physiology or circulation involves looking up systemic venous return directly to the pulmonary arterial tree (hence 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 etiology is unclear; however increased pulmonary venous pressures may contribute to affecting the respiratory epithelium and interruption of the bronchial mucus-causing parenchymal material to leak and hypersecretion of mucus with resultant cast formation. Histologic samples of patients with congenital heart disease reveal acellular casts with mucin affecting the respiratory epithelium and interruption of the bronchial mucosa causing inflammation. 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 revealsopacity and consolidated lung fields similar to pneumonia and,bronchoscopie or expectoration of a cast is an only reliable determinant of the diagnosis. See figure 2.

Nursing Care

The acute phase of illness requires care to be focused on protecting and maintaining a patient awake and alert by increasing high frequency ventilation with intermittent bronchoscopies performed to treat casts topically until the airway is clear. Oxygen therapy is often given long term supportive care, and development a medication regime that effectively prevents recurrence if possible. Patient and caregiver teaching are vital for patient safety and quality of life. Common upper respiratory illnesses or worsening congestive heart failure symptoms may confound the signs and symptoms of plastic bronchitis especially if the cough is non-productive. Education regarding chest physiotherapy techniques, maintaining a consistent medication schedule and prompt notification of their physician and healthcare team are of utmost importance. Frequent medical visits in the patient's community promotes prompt transfer to a tertiary center familiar with the disease process and capable of treating acute airway issues. Family caregivers must comprehend the need to intervene quickly when a cough and dyspnea develop by notifying their cardiologist as soon as the symptoms appear and information ought needs to be returned demonstrated by the "teach back" method to the nurse. Close monitoring of a recurrence is essential. Journals are helpful to patients and caregivers for tracking signs, symptoms, medication dosing and frequency, and the patient's general state of health. Important contact numbers should be listed in the front of the journal for easy access.

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Signs and Symptoms

The most common presenting symptoms are cough and intermitent dyspnea (Goldberg, Richly, & Dodds, 2010). The cough may be productive or non-productive depending on the patient’s ability to expectorate casts. Frequently, thick mucous is expectorated with a mucopurulent color that can be deveined as the mucus plug may actually be breaking away from the cast. Chest pain and fever are also reported but occurs 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 revealsopacity and consolidated lung fields similar to pneumonia and,bronchoscopie or expectoration of a cast is an only reliable determinant of the diagnosis. See figure 2.

Treatement

Once the airway is stabilized, patients with congenital heart disease suffering from the rare complication of plastic bronchitis are first evaluated for circulatory efficiency. A cardiac catheterization is performed to determine venous system pressures, development of collateral circulation in the lung, narrow pulmonary vessels and evaluation of the Fontan circuit. The Fontan conduit may be fenestrated to reduce systemic venous pressures; narrowed blood vessels may be stented, opened, collagenized, closed with obstructive devices and the functioning cardiac valve and ventricle assessed for optimum performance. Fontan physiology, at its best outcome mimics chronic, low grade congestive heart 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 dialated lymphatics, thoracic duct ligations are also considered especially if the patient exhibits problems with pleural effusions. (Colonari, Quarti, Pozzi, Gasparini, Carloni, & de Benedictis, 2014). Pharmacological therapy interventions include systemic and inhaled corticosteroids, mucolytics such as acetylcysteine and dextrose, sodium chloride, and fibrinolitics namely Urokinase and Alteplase. Tissue plasminogen activator (tPA) or Alteplase, is a serine protease that leads to localised fibrinolysis by converting plasminogen to plasmin. According to the literature topical application and nebulised tPA has been the most successful treatment without adverse effects in several case reports of children with congenital heart disease complicated by cast bronchitis. Brongen (et al., cited by Do et al., 2012) looked into the acute phase of illness and investigated use of saline, acetylcysteine and dornase alpha, and fibrinolytics namely Urokinase and Alteplase. The saline produced no effect, the Urokinase made the cast softer and, the sample treated with Alteplase, completely dissolved. See figure 3.

Figure 1 (a) Expectorated cast. (b) Cast extracted during flexible bronchoscopy. Copyright 2023 Hindawi Publishing Corporation.

Figure 2. Close monitoring of a recurrence is essential. Journals are helpful to parents and caregivers for tracking signs, symptoms, medication dosing and frequency, and the patient’s general state of health. Important contact numbers should be listed in the front of the journal for easy access.

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 after 15 minutes. Copyright 2012 American Academy of Pediatrics.

Conclusion


Additional Resources


Additional Resources