Plastic Bronchitis: A Worry No More

Abstract
Plastic bronchitis is a rare and mysterious condition. If at all recognized, it is well-known and characterized by the expectoration of bronchial casts. These bronchial casts obstruct airways and are life-threatening if undetected and untreated. Their uncommon nature makes them hard to diagnose as symptoms may mimic other respiratory conditions. Plastic bronchitis has been categorized into two types. One of which is easily treated and the other is not. A case study is discussed describing a young boy who developed plastic bronchitis after a Fontan surgery. His condition was resolved with the effective use of three medical imaging modalities during an embolization procedure of his lymphatic system.

Introduction
Plastic bronchitis is a rare pathologic condition in which casts form in the tracheobronchial trees of the lungs causing severe airway obstruction and asphyxiation (see Figure 1.A). Therefore, this condition will become life-threatening if undiagnosed and untreated. Not much is known about plastic bronchitis nor what triggers excessive secretion into the bronchioles. The condition can affect anyone at any age but mostly in children with congenital heart disease, especially after any corrective Fontan surgery procedures. The prevalence of plastic bronchitis in Fontan patients is estimated at 4-14%. Eberlein et al suggests that besides age, plastic bronchitis has a “female predominance.”

Plastic bronchitis can cause chronic coughing episodes that may result in the expectoration of airway plugs. When a cast is expectorated, these mucous plugs can resemble a small branch on a tree and act like a fingerprint of the airways (see Figure 1.B). Reports show that bronchial casts can vary in size, “from small segmental casts of a bronchus to casts filling the airways of an entire lung.” A large cast will cause pulmonary distress and certainly become life-threatening when major airways are obstructed. Casts will also have varying proportional combinations of cellular material consisting mainly of fibrin and mucin.
History

As unfamiliar as the medical world may be with plastic bronchitis, there are reports that describe similar conditions throughout history. The first known report of bronchial casts comes from Galen (A.D. 131 to 200) the famous Greek physician and philosopher whose studies have carved the way for much of our medical understanding even today. In the second century AD, he described a case of “venea arteriosea expectorati” and thought a cast was a collection of veins and arteries because of its appearance.\(^1\) Throughout time, recorded cases of bronchial casting has brought forth different names such as:

- Mucoid impaction.\(^1\)
- Fibrinous bronchitis.\(^1\)\(^-\)\(^4\)\(^,\)\(^7\)
- Pseudomembranous bronchitis.\(^1\)\(^-\)\(^4\)\(^,\)\(^7\)
- Hoffman bronchitis.\(^2\)\(^,\)\(^4\)\(^,\)\(^7\)
- Cast bronchitis.\(^4\)\(^,\)\(^7\)

The many names that are associated with this condition only add to the difficulty and challenge of identifying it.

Not only are there recorded instances during the course of history, there are also cases throughout the world documented from places such as Italy,\(^3\)\(^,\)\(^5\) Iran,\(^7\) Korea,\(^4\) Canada,\(^1\) and the United States.\(^2\)\(^,\)\(^8\)\(^-\)\(^9\) Therefore, plastic bronchitis cannot be linked to a lifestyle, culture, or ethnicity. Insight can also be gained from other countries. One of the unique qualities of plastic bronchitis is a flapping sound created from the vibrations of a large airway cast during respiration. In French it is called “bruit de drapeau” meaning the sound of a flag, and in German it is called “ventilgeräusch” meaning the sound of a fan.\(^1\)\(^-\)\(^2\) These are important higher lung sounds to be familiar with for the diagnosis of this condition as there are often no lung sounds at the bases.\(^8\)

Classification System

No one has been able to truly pinpoint the cause of bronchial cast formation nor how to classify them. For this reason, Seear et al\(^1\) studied nine well-documented cases of reported plastic bronchitis to create a classification system, and to, “provide a framework for further study of this obscure condition.”\(^\text{*}(p.364)\) They claimed that, “There is no accepted classification system for bronchial casts; but only a confusion of descriptive terms such as mucoid impaction,
During this same study a two-group classification system was created for identifying the differences in cast formation and composition. This system has become the most commonly referred to system for classifying plastic bronchitis casts to date.

The first group or type I is defined as the inflammatory group. It is characterized by its cast composition and by the inflammation of surrounding epithelium. These casts are composed mainly of fibrin with small amount of mucin, and are also associated with certain bronchial diseases with an acute presentation. The second group or type II is defined as the acellular group where, “cast production is chronic or recurrent.”2 It is characterized by the presence of hypocellular casts composed mainly of mucin (which are the mucous glycoproteins that compose the main components of airway mucus) with small amounts of fibrin. Type II casts seem to appear mostly in children with some form of heart disease, especially after any corrective surgery, which can be due to imbalances in pulmonary venous flow pressures causing leakage into the lungs. The theory is that trauma to the lymphatic vessels around the bronchus during corrective heart surgery and high venous pressure can cause leakage into the lungs and result in cast formation.

Detection and Diagnosis

Acute plastic bronchitis can be life-threatening and should be treated as urgent. Berlucchi et al supports this idea, “Knowledge of this disease is mandatory to perform correct diagnosis and provide prompt treatment.” Therefore, it is extremely important for all providers to know about and understand symptoms of plastic bronchitis. The unfamiliarity and rarity of this condition may cause patients to be incorrectly diagnosed, and many patients will go undiagnosed entirely. Healthcare providers have an extremely difficult task before them with any patient showing signs of respiratory distress. That difficult task is magnified if the patient shows signs of plastic bronchitis. Patients may present with a variety of symptoms with no true underlying cause. These may include symptoms such as:

- Acute respiratory distress.3
- Dyspnea.2-6,8,10
- Productive cough.2-6,8,10
- Chest pain.4,10
- Wheezing.\textsuperscript{2,6,8,10}
- Fever.\textsuperscript{3,6,8,10}
- Bronchial cast expectoration.\textsuperscript{5-6}
- Hypoxia.\textsuperscript{6}

Each patient can present with a different combination of these symptoms adding to the challenge of diagnosis. Clinical findings vary among patients depending on any associated illnesses as well as the production time of these lung casts. Some associated illnesses may include sickle cell disease,\textsuperscript{10} cystic fibrosis,\textsuperscript{8} asthma,\textsuperscript{4} and congenital heart defects.\textsuperscript{9}

The greatest key indicator is the expectoration of bronchial casts, although some patients are unable to have that experience. When patients are able to cough and expel these alien-like creations, they can take on varying sizes and shapes.\textsuperscript{1} Large casts, in particular, can mimic other diseases like tuberculosis and lung tumors (see Figure 3).\textsuperscript{1} Their rubbery consistency can vary depending on the proportions of fibrin, mucin, and other cellular material contained within each. Casts can display varying shades of beige\textsuperscript{6} and whitish-yellow\textsuperscript{2} colorations. One challenge in particular that health care providers may face is that a patient’s symptoms can mimic and be mistaken for aspiration of a foreign body.\textsuperscript{3} Eberlein et al\textsuperscript{2} records that even at times when casts are expectorated, they can be “mistaken for many objects, including foods such as noodles or chicken meat.”(p.165) In one particular instance parents believed their son had expectorated string cheese when it was in fact bronchial casts.\textsuperscript{6}

As might be expected, cast formations can be a secondary side effect to other diseases and conditions. In their study, Seear et al\textsuperscript{1} found similarities in the patients with acellular or type II casts. They all had hyper-secretions of airway mucus and some form of underlying congenital heart disease. This can create some confusion as excessive mucus production can easily be confused for several other diseases such as cystic fibrosis, chronic bronchitis, and severe asthma.\textsuperscript{1} Although plastic bronchitis may appear as a secondary condition, cast formation is not generally linked to other predisposing factors.\textsuperscript{1}

**Medical imaging**

It is well known that one of the most common medical imaging exams ordered is a chest x-ray. The reason for this is that a chest x-ray, as well as other medical imaging exams, can be a valuable tool in the visualization and diagnosis of diseases. Plastic bronchitis is no exception.
As described above, there are many symptoms for plastic bronchitis and radiologic findings can vary with each case. Berlucchi et al\(^3\) states, “The standard radiological evaluation of the chest may show atelectasic areas and compensatory hyperinflation in the contra-lateral side, whereas pneumomediastinum and emphysema are relatively frequent radiologic findings.”\(^{(p.205)}\) A thickening in the lower lobes without signs of an inhaled foreign body can also be a radiologic finding.\(^3\)

Each imaging modality will have its own contribution in a diagnostic effort. Computed tomography (CT) images can be used to aid in diagnosis, as well as, determine location of casts.\(^6\) Axial CT images of plastic bronchitis can be described visually as a “finger-in-glove” relationship when large airways are obstructed.\(^1\) Once plastic bronchitis is indicated, using various forms of medical imaging, a bronchoscopy may be performed to confirm findings in addition to treating the condition (see Figure 2). Not only can medical imaging be important for diagnosis, it can also be a vital tool in treating plastic bronchitis as is shown later in the case study.

**Treatment and Prognosis**

Like everything else in healthcare, treatment can vary greatly from patient to patient. There are two main approaches to treating patients with plastic bronchitis. First is to treat the condition itself and second is to treat the possible underlying cause. The best way to immediately alleviate the symptoms caused by plastic bronchitis is to remove the build-up within the airways. The first suggested approach is to perform various inhalation therapies\(^3\) but these are not generally successful. The best option is achieved by endoscopic extraction of the casts. Berlucchi et al\(^3\) stated, “Broncoscopy plus bronchial lavage is considered the gold standard therapeutic technique.”\(^{(p.204)}\) This can be a very difficult task as the casts are often too soft to grab and “too thick to be suctioned through a bronchoscope.”\(^{(p.165)}\) In some cases cryotherapy can be used to freeze the cast to aid in extraction.\(^2\) There are many challenges and risks to the patient during extraction of these casts as they may break apart and migrate further into the bronchus. It may be worth those risks because when extraction is successful it can provide some immediate relief for the patient.

While bronchoscopy may be an effective short-term treatment, understanding and treating the cause of the bronchial cast formation is the only way to provide a long-term...
treatment or even a possible elimination of this problem. As stated by Eberlain et al\textsuperscript{2}, “If plastic bronchitis presents as a complication of an underlying disorder, then optimal treatment of the underlying condition often leads to resolution of bronchial cast formation.”\textsuperscript{(p.167)} To treat the underlying condition providers need to pinpoint the type of cast formation their patient may have. Seear et al\textsuperscript{1} found that, “Survivors of type I casts seem to be well controlled with inhaled steroids.”\textsuperscript{(p364)} This isn’t the case with type II casts as “Steroids…are often found ineffective in plastic bronchitis caused by type II acellular casts.”\textsuperscript{2(p168)} Both of these observations have been confirmed by many others.\textsuperscript{1-2} Type II casts are not easily treated because they are generally a side effect to a congenital heart defect. By successfully treating the heart defect the condition of plastic bronchitis should resolve itself.

Prognosis varies depending on the type, cause, and patient situation. It is noted from Eberlain et al\textsuperscript{2} that, “Acellular casts generally carry a poorer prognosis than inflammatory casts, with inflammatory casts having a 6% to 50% mortality rate, whereas noninflammatory casts carry a 28% to 60% mortality rate.”\textsuperscript{(p.168)} Children who have type II casts due to congenital heart defects will struggle more because of the two conditions they must overcome. Seear et al\textsuperscript{1} stated, “Acellular bronchial casts carry a poor prognosis, probably reflecting the severity of the underlying cardiac disease.”\textsuperscript{(p.369)} They also observed that any attempt to address and treat type II casts themselves would be useless as they are the result of a complicated cardiac condition.\textsuperscript{1} Therefore, the best remedy for type II casts is to address and treat the cardiac disease of the patient. What this can mean to the patient is that if the cardiac condition cannot be resolved then the formation of bronchial casts will continue putting them at risk. Even though this doesn’t provide a lot of hope for those with type II casts the following case study is a shining beacon of what is to come in the new and advanced treatment of plastic bronchitis.

**Case Study**

A three-year-old boy with hypoplastic left heart syndrome underwent Extracardiac Fenestrated Total Cavopulmonary Connection which is a type of Fontan heart surgery. One of the complications the boy had from his surgery was chylous effusions or an excess of chyle fluid from the lymphatic system accumulating in the pleural cavity. At age five and a half he was admitted to a local hospital and presented with respiratory distress. During this visit, he was treated with antibiotics after being diagnosed with pneumonia. Two months later he began to
expectorate bronchial casts which led to a diagnosis of plastic bronchitis. He underwent a bronchoscopy procedure that revealed and confirmed casts in his right upper lobe caused by excessive fluid secretions. He was given various steroid and breathing treatments and then discharged from the hospital. Contrary to these treatments, he continued to produce bronchial casts.

At age six the boy was referred to a children’s hospital six months after being diagnosed with plastic bronchitis. At this institution, he was evaluated for a possible treatment for plastic bronchitis. He first had to undergo a non-contrast T2-weighted magnetic resonance (MR) lymphatic mapping. This showed significant dilation of the right peribronchial lymphatic ducts, as well as, supraclavicular and lumbar lymphatic networks (see Figure 4). Next, he underwent a contrast-enhanced dynamic MR lymphangiography. This difficult process involved bilateral ultrasound-guided needle punctures of the inguinal lymph nodes outside of the magnetic resonance imaging (MRI) scanner. After that, he was positioned into the scanner and simultaneous slow-hand injections of 2 mL gadolinium contrast and 2 mL of saline were placed into each lymph node over ten minutes during a time-resolved central k-space dynamic T1-weighted MRI. Sequence parameters were adjusted to capture images roughly every thirty seconds. This procedure showed a flow of the contrast starting at the dilated lumbar lymphatics and progressing towards the thoracic duct outlet. A dilated branch off the thoracic duct was observed as contrast progressed retrograde toward the carina and right lung hilum surrounding the airway (see Figure 5).

After discussing the imaging results and a possible lymphatic embolization procedure with the parents, a decision was made to try more aggressive therapies and breathing treatments first. The patient’s symptoms only improved slightly over the next three weeks and so he returned to the same children’s hospital. A new decision was made to attempt an interventional procedure to resolve his plastic bronchitis. A team assessed the previous MRI scans and determined the probable cause of the bronchial casts was the dilated branch off the thoracic duct that allowed retrograde flow. This was the cause of peri-bronchial lymphatic congestion. It was determined that this lymphatic congestion causes a “proteinaceous fluid” leak into the airway on the right side. After careful consideration, they created a plan and went forth with this complex procedure.
Under general anesthesia, once again, ultrasound-guided needle punctures were initiated into the left and right inguinal lymph nodes. Injection of 2 mL of Lipiodol (an ethiodized oil contrast agent) was then administered to aid in viewing correct positioning. Utilizing fluoroscopic guidance, an anterior trans-abdominal approach was taken using a 22-gauge Chiba needle into the cisterna chyli. Then a guidewire was placed into the thoracic duct and a microcatheter was advanced over the wire to inject contrast into the thoracic duct. The contrast helped to confirm the MRI findings as it revealed once again the lymphatic branch that progressed to the right hilum. The microcatheter was then carefully positioned into the branch and embolization was initiated and confirmed with 4 mL of more Lipiodol contrast (see Figure 6). The last step was to then inject 1 to 2 mL of n-Butyl cyanoacrylate in the proximal part of the lymphatic branch. This substance works as a glue closing off the flow through that lymphatic vessel. Other important vessels were occluded beforehand to prevent any improper flow of contrast or glue during the procedure.

It was reported that the boy recovered from the procedure without complications and was discharged three days later. He did have chest pain after two days that resolved, and he was able to discontinue his breathing treatments and respiratory therapies after two weeks. It was also reported that after five months he no longer had any symptoms of plastic bronchitis, “free of coughing spells and casts.”

**Discussion**

In this case, there is a great example of successful treatment of a patient with type II cast formation. The medical professionals recognized that the cast formation was an underlying symptom of retrograde lymphatic flow, and they were able to isolate it and treat it effectively. It is also observed that ultrasound, MRI, and fluoroscopy were utilized during all aspects of this young boy’s case. Successful diagnosis and treatment would not be possible without the use of medical imaging. The medical staff understood and effectively utilized ultrasound, MRI, and fluoroscopy. This case portrays the importance of teamwork in any medical imaging department.
Conclusion

Plastic bronchitis is a dangerous condition that causes severe respiratory distress and is often fatal.\textsuperscript{9} It is a difficult condition to diagnose because it can be mistaken for other respiratory conditions and therefore be mistreated. Seear et al\textsuperscript{1} claims that bronchial casts are becoming increasingly rare as general health improves. So far, the biggest danger may be the lack of knowledge and understanding healthcare providers may have. Eberlein et al\textsuperscript{2} indicates that a registry of patients with plastic bronchitis has been initiated to help health care providers learn and evaluate different treatment options. This is a great process since some evidence suggests that part of the problem is that many cases go underreported.\textsuperscript{6} Type I is effectively treated with steroids while type II needs to be addressed at the underlying cause. Through their courage and success, Dori et al\textsuperscript{9} has opened doors to new ways of treating the cause of plastic bronchitis. As our knowledge and treatment possibilities increase, plastic bronchitis can be effectively diagnosed, treated, and become a worry no more.
References


Figures and Captions

Figure 1. A. Superior view of a bronchial cast in right upper lobe from a bronchoscopic procedure showing airway obstruction. B. Image of a large bronchial cast once it is extracted. This particular cast is large enough to show the middle and right lower lobe divisions. Images courtesy of: Eberlein MH, Drummond MB, Haponik EF. Plastic bronchitis: a management challenge. *Am J Med Sci.* 2008;335(2):163-169.
Figure 4. Images of the T2-weighted MRI scan showing the dilated network of peribronchial lymphatic channels on the right side as indicated by the arrow. (Arrowheads point to dilated supraclavicular and abdominal lymphatic networks.) B. The airway is shown in green to help portray the relationship between the right bronchus and lymphatic network of interest. Images courtesy of: Dori Y, Keller M, Rychik J, Itkin M. Successful treatment of plastic bronchitis by selective lymphatic embolization in a Fontan patient. *Pediatrics* [serial online]. August 2014;134(2):e590-e595. Available from: MEDLINE, Ipswich, MA. Accessed October 31, 2014.
Figure 5. A-C. Images of MR lymphangiogram during three different times in the study (shown in seconds in top left). A. Showing contrast in dilated lumbar lymphatic vessels. B. Arrowhead indicating contrast passed cisterna chyli. Arrows pointing to the lymphatic branch of interest as contrast moves through the dilated branch to the left, then upward, and then right. C. Boxed area shows that contrast has arrived into the dilated peribronchial lymphatic network and surrounds the right bronchus. Arrowheads pointing to left supraclavicular lymphatics. D. Schematic of the lymphatic anatomy. Black arrows indicate direction of lymph flow. Images courtesy of: Dori Y, Keller M, Rychik J, Itkin M. Successful treatment of plastic bronchitis by selective lymphatic embolization in a Fontan patient. Pediatrics [serial online]. August 2014;134(2):e590-e595. Available from: MEDLINE, Ipswich, MA. Accessed October 31, 2014.
Figure 6. A-D. Images of the lymphangiogram procedure using fluoroscopy. A. Seven seconds after contrast injection inferior to cisterna chyli showing contrast movement into the dilated branch of interest confirming previous MRI study. White arrows showing direction of flow and arrowhead showing catheter tip. B. Fifteen seconds after contrast injection as it fills the lymphatic network surrounding the right bronchus. C. Arrowhead showing site of catheter embolization of the lymphatic branch of interest as contrast is injected again to verify positioning. D. Arrow showing glue filled in proximal end of lymphatic branch of interest as contrast continues to fill the end of the dilated peribronchial lymphatic vessels (box), thus, ending the procedure. Images courtesy of: Dori Y, Keller M, Rychik J, Itkin M. Successful treatment of plastic bronchitis by selective lymphatic embolization in a Fontan patient. Pediatrics [serial online]. August 2014;134(2):e590-e595. Available from: MEDLINE, Ipswich, MA. Accessed October 31, 2014.