

SUCCESSFUL TREATMENT OF PLASTIC BRONCHITIS WITH LOW FAT DIET AND SUBSEQUENT THORACIC DUCT LIGATION IN CHILD WITH FONTAN PHYSIOLOGY

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ABSTRACT

Plastic bronchitis is a rare condition characterized by the formation and expectoration of long, branching bronchial casts that develop in the tracheobronchial tree and cause airway obstruction. Plastic bronchitis has become increasingly recognized as a feared complication of the Fontan operation with a mortality of up to 50%. We report an 11 year old boy who developed severe plastic bronchitis following Fontan repair and the successful long-term control of cast formation utilizing a low-fat diet and subsequent thoracic duct ligation.

Keywords: plastic bronchitis, Fontan procedure, chyloptysis, low fat diet, thoracic duct ligation, lymphoscintigram

CASE REPORT

An 11-year-old boy with a history of hypoplastic left heart syndrome and multiple surgical palliations presented with increasing respiratory symptoms. His surgical history consisted of a Norwood procedure at 8 days of life (neo-aortic construction utilizing native pulmonary artery and creation of aorto-pulmonary or B-T shunt for pulmonary blood

flow), a bidirectional Glenn at 4 months (superior vena cava to pulmonary artery anastomosis and ligation of B-T shunt) and a lateral tunnel fenestrated Fontan (inferior vena cava baffled through right atrium and anastomosed to pulmonary artery) with tricuspid valvuloplasty at 22 months of age. At 4 years of age he was noted with worsening tricuspid regurgitation warranting tricuspid annuloplasty and closure of the fenestration. He had a family history of congenital heart disease.

He was asymptomatic until age 10 when he was diagnosed with asthma and experienced exacerbations of increasing frequency. By 11 years of age he was experiencing frequent (1-2 times/week) violent, post-tussive expectoration of malleable, white bronchial casts (*Fig. 1*) and required supplemental oxygen to maintain saturations in the 80%'s. Histopathologic and chemical analysis determined the casts were acellular and lymphatic in origin, containing triglycerides, chylomicra, and foamy macrophages with oil-red-O positive lipid droplets (*Fig. 2*).

A cardiac catheterization was performed which revealed favorable Fontan hemodynamics with a mean Fontan pressure of 12mmHg and a normal left atrial pressure of 8mmHg. Multiple medical treatment

strategies were attempted including albuterol, inhaled alteplase, inhaled tissue plasminogen activator (tPA), and low-molecular weight heparin subcutaneous injections with minimal benefit. Cardiac medications included angiotensin converting enzyme (ACE) inhibition. He had a previous history of post-operative atrial flutter but cardiac monitoring revealed sinus rhythm without sustained atrial arrhythmias. Nevertheless, an epicardial atrial pacemaker was placed to eliminate sick sinus syndrome as a possible contributing factor.

The patient was hospitalized nearly continuously for a year with profound respiratory symptoms and almost daily cast production, requiring multiple therapeutic bronchoscopies to remove obstructive casts. A continuous octreotide infusion of 5mcg/kg/hr achieved initial success but within two months, his symptoms and cast production returned and proved unresponsive to octreotide. At that time, bipedal ^{99m}Tc sulfacolloid lymphangioscintigram revealed normal peripheral and central lymphatic transport but abnormal radiotracer reflux at the level of the mid-thoracic duct into the tracheobronchial tree and right upper lung field (*Fig. 3*). A conventional oil contrast lymphangiogram was attempted to further pinpoint the site of chylous lymph reflux but no suitable pedal lymphatic could be found for infusion. CT revealed racemose low flow vessels in the right axilla. His diet was changed from a low-fat diet with only fair compliance to a staunch no-fat diet. Intravenous lipid (Intralipid 20%) was infused weekly and overnight G-tube feeds with Carnation® instant breakfast juice was initiated to meet 50% of his daily caloric needs. In order to meet essential fatty acid requirements, 2-3% of the total calories included medium chain fatty acids, and fatty acid plasma levels were monitored. With this fastidious dietary regimen, his cast production ceased, and he was quickly able to be weaned off supplemental oxygen. His plastic bronchitis was thus controlled for 3¹/₂ years as an out-patient, and he returned to school full time.

As a teenager, his compliance with a modified low fat diet waned, and he had several lapses with fatty food consumption, which were associated with rapid cast production, necessitating hospitalization for oxygen and pulmonary hygiene for symptomatic relief. Based on the findings of thoracic duct lymph reflux on the earlier lymphangioscintigram and his poor adherence to a low fat diet with a return of debilitating symptoms, a thoracic duct ligation was entertained as a potential means to halt the direct spill of chyle into the tracheo-bronchial tree and lungs.

Following several conferences with patient, family, and his team of caregivers including cardiologist, pulmonologist, lymphologist, and cardiothoracic surgeon, the procedure was successfully performed at age 15. The surgery was preceded by therapeutic bronchoscopy to maximize airway patency. A right thoracotomy incision was made at the level of the 8th intercostal space entering the right hemithorax. Working anterior to the spine, a large, distended clear fluid-containing thoracic duct was noted posterior and to the right of the esophagus. A suture was placed to encompass all tissue posterior to the esophagus from aorta to the right side of the spine including the thoracic duct. During the recovery period, cast production briefly resumed but appeared to respond to high dose afterload reduction (Captopril 25 mg three times a day). Within 2 weeks of surgery, the patient was able to tolerate a regular diet, and he has remained cast-free ever since. He did develop bilateral chylous pleural effusions several months post-operatively, which required drainage and pleurodesis.

Following the thoracic duct ligation, he experienced a rapid growth spurt and was noted to have increased tricuspid stenosis, which required a tricuspid repair approximately one year following his thoracic duct ligation. He did well post-operatively without evidence of plastic bronchitis recurrence.

Today at age 17, two years following

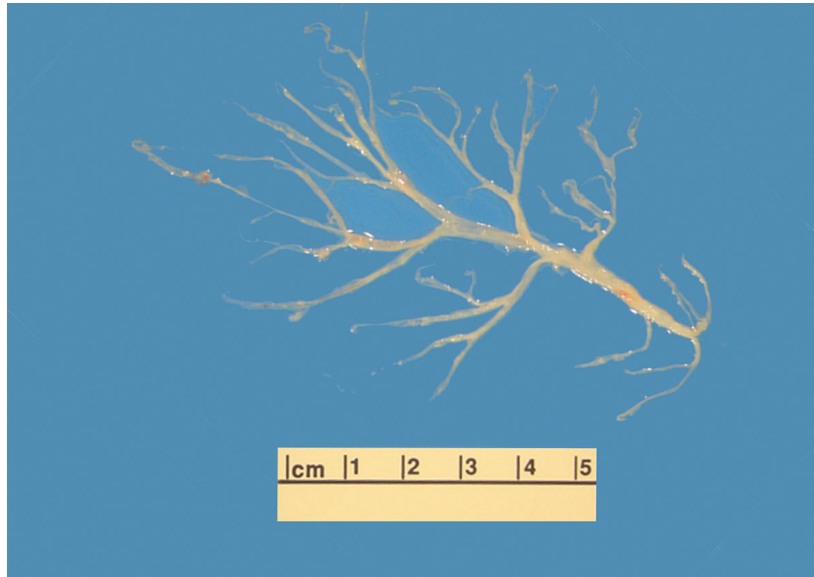


Fig. 1. Large arboreal bronchial cast preserved following expectoration.

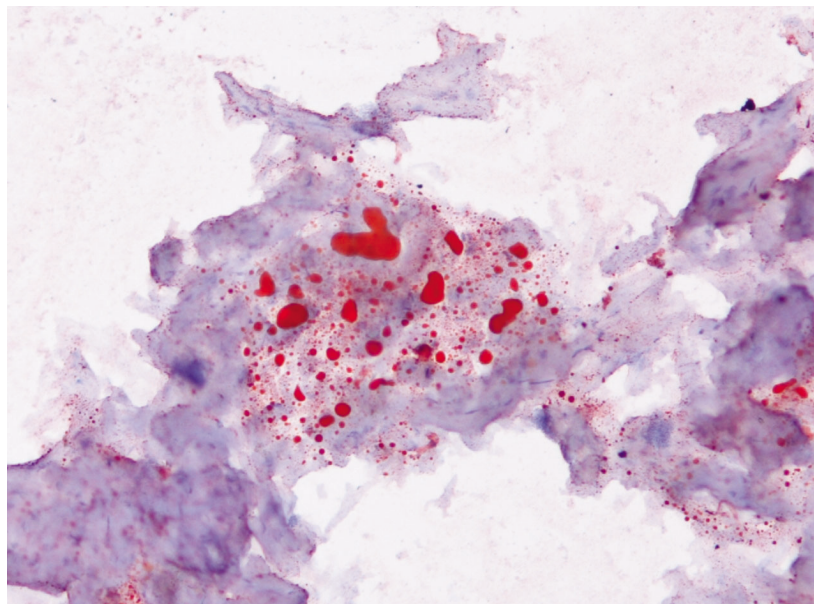


Fig. 2. Acellular cast with oil red O positive lipid droplets, original magnification X200.

thoracic duct ligation, he continues on diuretics and ACE inhibition, but remains cast- and symptom-free, on a regular diet without fat restriction and is attending high school.

DISCUSSION

Plastic bronchitis is a rare condition characterized by the formation and expectoration of long, branching bronchial casts that develop in the tracheobronchial tree and cause airway obstruction (1).

The first descriptions of Plastic Bronchitis

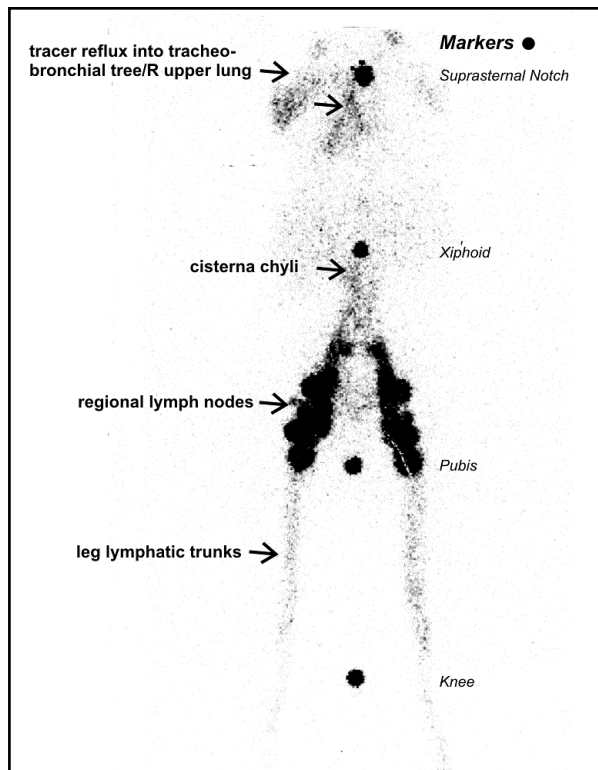


Fig. 3. Bilateral lower extremity ^{99m}Tc sulfacolloid lymphangioscintigram. Note prompt cephalad tracer transport from foot injection sites into peripheral and central lymphatic trunks and regional inguinal and retroperitoneal lymph nodes up to the level of the cisterna chyli with demonstrable retrograde reflux of tracer (arrow) into the tracheobronchial tree and right upper lung field. Radioactive markers placed at key anatomic landmarks – knee, pubis, xiphoid, and suprasternal (SS) notch.

likely date back to AD 131-200 with Galen's description of "venae arteriosae expectorantii." The characteristic branching, bronchial casts have subsequently been referred to as fibrinous bronchitis or pseudomembranous bronchitis and most recently, plastic bronchitis in the 20th century (2). Plastic bronchitis has become increasingly recognized as a feared complication of the Fontan operation with a mortality of up to 50% (3).

Sear et al proposed a two-type classification: type 1 consisted of inflammatory casts associated primarily with bronchopulmonary disorders and type 2 or acellular casts associated with cyanotic congenital cardiac disorders (2,3-5). A more recent classification by Madsen et al segregates by underlying disease (Congenital Heart Disease,

Asthma, Lymphangiectasis and Sickle Cell disease) plus cast histology (including mucinous, fibrinous, inflammatory and chylous) and recognizes that patients with congenital heart disease carry the highest risk of mortality from plastic bronchitis (6).

Various congenital heart lesions have been associated with the development of plastic bronchitis. Fontan physiology, the ultimate result of single ventricle surgical palliation, in which both the inferior and superior vena cavae are connected directly to the pulmonary arteries, has increasingly become recognized as a risk factor for plastic bronchitis (4,5,7-10).

A Fontan circuit flows passively, without the pumping force of the right ventricle, and therefore requires a high central venous

pressure (CVP) to drive pulmonary blood flow. In a normal heart, the mean CVP is 0-5 mmHg, whereas in a well functioning Fontan circuit, CVP is typically 10-15 mmHg. The lymphatic circulation drains directly into the systemic venous system and therefore an obligatory increase in lymphatic pressure from resistance to lymph flow occurs with a Fontan connection. Additionally, hepatic and intestinal (as well as more general) congestion associated with increased systemic venous pressure directly contributes to an increase in lymph flow into the thoracic duct (11,12). The combination of increased thoracic duct lymph formation with elevated lymphatic pressure likely provides the ideal environment for development of abnormal lymph-tracheo-bronchial-alveolar fistulae in the Fontan circulation (13,14). Anomalous lymphatic connections may also coexist congenitally in patients with complex congenital heart disease thus providing a preexistent substrate for bronchopulmonary connections. Indeed, rapid development of casts within days following a Fontan operation has been reported (13). Thus the Fontan circulation appears to provide a uniquely opportune setting for the development of the chylous form of plastic bronchitis precipitating chyloptysis.

During the work-up for plastic bronchitis in the patient with Fontan physiology, a thorough hemodynamic evaluation should be performed including echocardiography, cardiac catheterization and rhythm evaluation. Aberrations that result in elevated systemic and/or pulmonary venous pressure in the Fontan circuit include arrhythmias, AV valve regurgitation, poor single ventricle function, elevated pulmonary vascular resistance, and significant aorto-pulmonary artery collateral vessels. Resolution of plastic bronchitis has been reported following interventions that result in improved Fontan hemodynamics such as the re-establishment of A-V synchrony via atrial pacing and following Fontan fenestration (7,15). A thorough cardiovascular evaluation in our patient failed to reveal significant hemodynamic aberrancies, and

thus we focused our efforts on cast identification and treatment.

Various treatment strategies for plastic bronchitis have been attempted, typically with limited success. Most therapies serve to soften the casts and aid non-obstructive expectoration and include: various nebulization treatments, systemic and inhaled corticosteroids, tPA, urokinase, and DNase (3,4,8,9). Serial bronchoscopies to facilitate cast removal have been explored as potential treatment option, however the unusually friable nature of large bronchial casts poses a challenge (16). Pulmonary lobectomy to relieve severe hypoxemia secondary to casts extending from the right middle lobe has also been reported (16).

Recognition of the lymph composition of the casts and demonstration of the abnormal refluxing lymph circulation on lymphscintigram were important in our case as we focused on interventions designed to decrease chyle production and flow. We initially attempted a low-fat diet and noted temporal resolution with octreotide, a somatostatin receptor stimulant, which reduces lymph flow by increasing splanchnic arteriolar resistance and decreasing gastrointestinal blood flow (17) although an additional direct effect on the lymphatics has not been investigated. Following return of cast production, we placed the patient on a staunch non-fat diet without octreotide and noted rapid cast resolution.

Previous studies have proposed dietary modification with elimination of long-chain triglycerides for the management of plastic bronchitis (4,17,18), but to our knowledge, this is the first pediatric report that clearly demonstrates a non-fat diet even alone as a successful treatment option for this condition. Strict long-term compliance with a low fat diet is challenging and ultimately proved to be the limiting quality of life factor in our teenage patient. Therefore, a thoracic duct ligation was performed. Thoracic duct ligation has been previously described with varying degrees of success (4,5,19). Our successful surgical outcome and prior

management was positively influenced by performing early minimally invasive (bipedal intradermal injection of radiotracer) lymphoscintigraphy, which clearly delineated the central lymph-bronchial fistulous connections and directed the general management and ultimate surgical approach proximal to the site of reflux.

In conclusion, we report the successful treatment of a life-threatening and protracted case of plastic bronchitis in a child with Fontan physiology. After a thorough hemodynamic evaluation failed to reveal aberrancies, a systematic multidisciplinary team approach involving cardiac, pulmonary, pathology, lymphatic, imaging, and surgical specialties was utilized. With recognition of the intestinal lymph composition of the bronchial casts, treatments directed at reducing chylous flow were initiated, and success was achieved with a non/low fat diet and subsequent thoracic duct ligation.

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