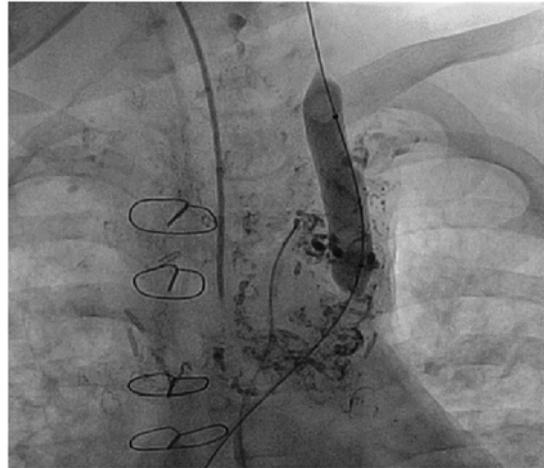


Novel Treatment for Post-Fontan Plastic Bronchitis by Direct Lymphatic Embolization

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Introduction: Plastic bronchitis is a rare, potentially life-threatening complication of Fontan procedure for single ventricle palliation. For these patients, the 5 year mortality rate is as high as 50%, increasing dramatically if the diagnosis is made less than one year post surgery. Current conservative treatment includes frequent bronchial lavage, anti-heart failure medications, anti-pulmonary hypertension medications, anticoagulation, and tissue plasminogen activator, DNase, N-acetyl cysteine, and hypertonic saline inhalations. Recent publications suggest selective lymphangiography and embolization as a promising new treatment option with good short term outcome.

Methods: We present a case of a 3.5-year-old boy with double inlet left ventricle, malposed great arteries, who developed severe plastic bronchitis 4 months post his Fontan completion. After recurrent life threatening episodes and failure of 8 month of conventional therapy, the patient was referred for lymphatic catheterization with direct embolization. Intranodal lymphangiogram was performed with injection of Lipiodol® (ethiodized oil-based contrast agent) into the inguinal lymph nodes. Under fluoroscopic guidance, the cisterna chyli was fluoroscopically accessed via anterior transabdominal approach. A guidewire and microcatheter were advanced into the thoracic duct and manipulated towards distal branches. Methylene blue injection into the lymphatic system resulted in observed staining of the right bronchial tree on simultaneous bronchoscopy. Embolization of distal lymphatic collaterals was performed by Lipiodol injection in the distal lymphatic vessels, followed by coil embolization of the thoracic duct and injection of glue.

Results: Post procedure the patient required mechanical ventilation and inotropic support for several days. He developed bilateral pleural effusions and moderate ascites that resolved with conventional treatment with no need for drainage. He was discharged home 18 days after the procedure. Currently, 2 months post intervention he is free of plastic bronchitis casts, only on anti-cardiac failure and anti-pulmonary hypertension medications.

Conclusions: Direct lymphatic embolization may be an effective novel treatment for plastic bronchitis with excellent short term prognosis.

Figure: Selective lymphangiography demonstrating abnormal dilated thoracic lymph vessels. These vessels were occluded using Lipiodol® injection with simultaneous balloon inflation in the left superior vena cava to avoid contrast embolization into the pulmonary tree.