CASE REPORT

Transvenous retrograde thoracic duct embolization for effective treatment of recurrent plastic bronchitis after Fontan palliation

Raymond N. Haddad MD1 | Raphael Dautry MD2 | Damien Bonnet MD, PhD1,3 | Sophie Malekzadeh-Milani MD1

1M3C-Necker, Hôpital Universitaire Necker-Enfants Malades, Assistance Publique-Hôpitaux de Paris (AP-HP), Paris, France
2Department of Radiology, Hôpital Cochin, Assistance Publique-Hôpitaux de Paris (AP-HP), Paris, France
3Université de Paris Cité, Paris, France

Correspondence
Raymond N. Haddad, MD, Service de cardiologie congénitale et pédiatrique, Centre de référence des malformations cardiaques congénitales complexes—M3C, Hôpital universitaire Necker-Enfants malades, 149, rue de Sèvres, Paris 75015, France.
Email: raymondhaddad@live.com

Abstract
We report the case of a 5.5-year-old patient (16 kg/105 cm) who presented with plastic bronchitis (PB) refractory to conservative treatment 3 months after completion of Fontan palliation. Bi-inguinal transnodal fluoroscopy-guided lymphangiogram confirmed the chyous leak originating from the thoracic duct (TD) into the chest and did not opacify any central lymphatic vessel for direct transabdominal puncture. Retrograde transfemoral approach was adopted to catheterize the TD and selectively embolize its caudal portion using microcoils and liquid embolic adhesive. Recurrence of symptoms after 2 months indicated a redo catheterization to occlude the TD entirely using the same technique. The procedure was successful and the patient was discharged after 2 days with sustained clinical improvement at 24 months postoperative. In the context of refractory PB, end-to-end transvenous retrograde embolization of the TD appears to be an interesting alternative to more complex interventions such as transabdominal puncture, decompression, or surgical ligation of the TD.

KEYWORDS
bronchitis, congenital, Fontan circulation, heart defects, lymphography, thoracic duct

1 | INTRODUCTION

Plastic bronchitis (PB) is a rare but potentially detrimental complication of the Fontan circulation.1 Understanding of this disorder has improved over the past years with the advances in lymphatic system imaging and encouraging outcomes of lymphatic transcatheter interventions.1 Thoracic duct embolization (TDE) has been proposed as a treatment strategy for lymphatic dysfunction.1–4 We report an interesting pediatric case of PB relapse 2 months after retrograde transvenous embolization of the caudal end of the thoracic duct (TD) and requiring end-to-end TDE for complete and sustainable resolution of symptoms.

2 | CASE PRESENTATION

A 5.5-year-old male patient (16 kg/105 cm) was referred to our institution for severe recurrent PB. He had a prenatal diagnosis of unbalanced atrioventricular septal defect with left heart hypoplasia and was palliated with pulmonary artery banding at 1 week followed...
by Damus-Kay procedure with right Blalock-Taussig shunt and aortic arch enlargement at 6 weeks. Glenn procedure was done at 4 months of age after balloon dilatation of the aortic arch. Fenestrated Fontan procedure was completed at 5 years with an uncomplicated postoperative course. The patient was admitted to the pediatric intensive care unit 3 months later for acute respiratory distress. Computed tomography scan showed multiple bilateral areas of patchy consolidation and ground-glass opacity with obstruction of the right main and right lower bronchi. He was diagnosed with PB based on expectorated airway casts. Cardiac catheterization showed elevated central venous pressure at 20 mmHg with left atrial pressure at 9 mmHg and a patent fenestration. Oxygen saturation in the descending aorta was measured at 88%, with a mixed venous saturation of 68%. Two large aortopulmonary collaterals were embolized with a drop of the central venous pressure to 16 mmHg. There were no other residual hemodynamic lesions requiring catheter-based correction. Despite aggressive conservative therapy, he continued to have frequent casts and was referred for TDE. Under ultrasound guidance, bilateral inguinal lymph nodes were directly accessed using a 22-G/9 cm fine needle. Lymphangiography was performed under intermittent fluoroscopy guidance with ethiodized oil (Lipiodol® Ultra fluide; Guerbert) to determine the anatomy and flow pattern of lymph in his central lymphatic system (Figure 1A). During infusion, serial spot radiographs did not opacify a cysterna chyli or another target central lymphatic vessel obviating the possibility of a direct transabdominal TD needle access. Diffuse and abnormal lymphatic leak into the chest, predominantly right-sided was observed and the junction of the cervical part of TD with the left brachiocephalic vein was located (Figure 1B, Supporting Information: Video 1). The TD venous ostium was retrogradely cannulated from the right femoral vein using a telescoping combination of a 4Fr Judkins Left 2.0 diagnostic catheter (Cordis Corp.), 2.7Fr Progreat™ microcatheter system (Terumo Corp), and a 0.014-in. Fathom™ steerable guidewire (Boston Scientific Corp.) (Figure 1C, Supporting Information: Video 2). Selective ductography with iodinated contrast showed a looking-like type 2 lymphangiography pattern with a proliferative peribronchial lymphatic network, branching off the TD, and progressing mainly toward the mediastinum and the right lung hilum (Figure 2, Supporting Information: Video 3). These vessels were numerous, too small, and tortuous to be occluded individually. After flushing with G5 solution, the lower third of the TD was sealed using two 3D Concerto™ coils (Medtronic) and a

**FIGURE 1** Bi-inguinal transnodal lymphangiography (A) showing predominant right-sided contrast extravasation in the chest (white circle) and localizing the lymphovenous junction near the internal jugular and subclavian vein confluence (white pointed arrow) (B). Transvenous retrograde cannulation of the thoracic duct (C).

**FIGURE 2** Selective ductography showing proliferative and dilated paratracheal lymphatic ducts.
1/1 mixture of ethiodized oil (Lipiodol® Ultra fluide; Guerbert)/N-butyl-2-cyanoacrylate metacrilosisolfolane glue (Histoacryl® L, B. Braun Surgical) (Figure 3, Supporting Information: Video 4). A redo catheterization was performed after 2 months for recurrence of PB and revealed the persistence of dilated peribronchial lymphatic network with a residual leak across the previously embolized caudal section of the TD (Figure 4A,B, Supporting Information: Video 5). The central venous pressure was 15 mmHg. We did not identify lymphovenous valve incompetence during left venous angle contrast interrogation. Embolization of the TD was performed over its entire length with intercalation of glue and three Concerto coils (Figure 4C, Supporting Information: Video 6). Occlusion of the cervical part of the TD was performed with coils to obviate the risk of glue embolization into the venous circulation. The patient was rapidly discharged. Sustained clinical improvement without subsequent bronchial cast recurrence was confirmed after 24 months of follow-up. The patient did not report any TDE-related complications such as chronic leg swelling, abdominal swelling, or chronic diarrhea. The patient was kept under sildenafil and spironolactone therapy and has an oxygen saturation of 95%. Last Doppler ultrasound showed a respiratory variation of the caval flow and low-velocity (0.3–1.4 m/s) fenestration flow.

3 | DISCUSSION

PB occurs in almost 4% of patients with total cavopulmonary connections.1 Fontan circulation physiology chronically increases systemic venous pressure, resulting in a higher afterload at the site of the TD outflow, which impedes effective drainage of the lymphatic system. This disorder also involves elevated lymphatic production and in some patients local abnormalities of the lymphatic system. The double burden of increased lymph production in the system in which drainage is already impeded by venous hypertension is likely one of the main mechanisms of lymphatic dysfunction. This results in abnormal lymphatic collateralization, overdistention, and rupture resulting in lymphatic extravasation into different low-pressure lumens such as the airways as seen in our patient.1,5,6

Percutaneous lymphatic interventions are well-established interventional radiology procedures and are considered less invasive alternatives to surgical treatment of traumatic, postsurgical, malignant, or iatrogenic chylous leaks.2,3 Likewise, different percutaneous TD interventions have been reported in patients with congenital heart disease and lymphatic failure (Table 1).4,7–16 TDE requires a comprehensive understanding of the anatomy of the TD and cisterna chyli as well as their anatomical variations. In our patient, the lymphangiogram did not identify a target central lymphatic vessel for the percutaneous transabdominal approach.4,7,9,11 Direct cervical puncture could have been an alternative but it was not considered the best option in this pediatric case.17 As the TD venous junction was depicted, we decided to access the TD via retrograde transvenous catheterization.18,19 A limitation of this technique is that the feasibility depends both on the visualization and on the anatomy of the TD cervical part and lymphovenous junction. In cases where this junction is either not seen or plexiform, retrograde access may be impossible.11 It can be argued that passing across the lymphovenous valve might impair its function in regulating the lymph flow into the venous circulation. Manipulation of low-profile material by an experienced radiologist was important to mitigate this risk of iatrogenic valve incompetence that might worsen the condition and facilitates retrograde blood entry from the high-pressure circulation into the low-pressure lymphatic system. Superselective retrograde embolization of the transected lymphatic branches and isolation of the TD with covered stenting have been reported to exclude the retrograde flow into the airways.4,18 Both of these interventions can offer important advantages over end-to-end TDE such as saving the sequelae of embolizing the entire TD. Maintaining TD patency can be important in patients with significant lymphatic congestion and targeted treatment of only the pathologic lymphatic vessels can potentially improve the outcome of such interventions.
Another viable treatment option is the disruption of cisterna chyli or the decompression of the TD into a lower-pressure chamber but both of these procedures are risky and can be technically challenging. In our patient, abnormal lymphatic vessels were small and numerous and were originating at different levels from the TD. This anatomy was considered unfavorable for either selective embolization of all pathological lymphatic vessels or stenting of the TD as it would have required the implantation of several overlapping covered stents. We chose to first embolize the caudal TD because we believed it would be sufficient to eliminate TD flow toward the lungs while mitigating the risk of the procedure. Embolization of the caudal TD has not been always reported as successful in the setting of a transected TD. Likewise, this technique was not clinically beneficial for the patient. The recurrence of PB was probably due to the incomplete closure of the TD caudal portion and the collateral pathways draining into the proximal portion beyond the area of caudal embolization with backflow into the leaks. Therefore, we decided to entirely occlude the TD to eliminate any lymph inflow to the lungs, and the outcomes were very satisfactory. This minimally invasive approach allowed us to avoid more complex lymphatic interventions such as transabdominal puncture, TD decompression, or surgical ligation.

When planning for such minimally invasive lymphatic procedures, it is essential to consider possible associated risks and collaborate with experienced interventional radiologists. The use of an oil-based contrast agent in children with right-to-left shunting poses a risk for stroke and fatty pulmonary emboli. Occlusion of right-to-left shunts has been performed to minimize that risk. Transvenous balloon occlusion of the lymphovenous junction is also an option. During the first intervention, we occluded the TD very distally minimizing that risk. During the second intervention, we did not perform temporary balloon occlusion of the fenestration during the lymphangiogram because the high central venous pressure was considered “protective” against the glue backflow from the lymphatic system into the venous circulation. The risk of glue migration into the venous circulation was also mitigated by sealing the cervical part of the TD with coils and by minimizing the amount of contrast agent, which is of utmost importance. The lymph system is generally a low-pressure system that responds to subtle pressure differences. Delayed functional complications related to TDE are conceivable because of the intentional redistribution of lymphatic flow dynamics induced by the procedure. A previous study reported a 14.3% rate of probably related long-term complications after technically successful TDE. Therefore, longitudinal follow-up is required to further define the long-term effect of this treatment. It is important to consider that TDE does not address the underlying cause of this PB, which is significant lymphatic congestion due to the Fontan physiology of elevated venous pressure. We have continued medical treatment to encourage pulmonary vasodilation and optimize cardiovascular circulation, thereby reducing ongoing lymphatic complications.
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<td>Hubrechts et al.(^{15})</td>
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</table>

Abbreviations: CS, coronary sinus; CTA, computed tomography angiography; DCMRL, dynamic contrast-enhanced magnetic resonance lymphangiogram; FU, Follow-up; IL, intranodal lymphangiography; IV, innominate vein; LA, left atrium; L SVC, left superior vena cava; MRI, magnetic resonance imaging; NOP, number of patients; PB, plastic bronchitis; PLE, protein-losing enteropathy; Pt, patient; QOL, quality of life; TD, thoracic duct; TDD, thoracic duct decompression.
4 CONCLUSION

We demonstrate that the etiology of recurrent PB in a child with Fontan physiology was retrograde flow from the TD into peribronchial lymphatic collateral networks. Fluoroscopy-guided lymphangiography was essential to understand the anatomy and flow pattern of the lymphatic system and to plan the lymphatic intervention. End-to-end TDE is an effective treatment in Fontan patients with PB and can be necessary in case of relapse after selective embolization of the TD caudal section. This minimally invasive retrograde transvenous approach is an interesting treatment alternative to more complex lymphatic interventions.

CONFLICT OF INTEREST STATEMENT

The author declare no conflict of interest.

ORCID

Raymond N. Haddad https://orcid.org/0000-0001-7605-4708
Damien Bonnet https://orcid.org/0000-0002-8722-5805
Sophie Malekzadeh-Milani https://orcid.org/0000-0002-9527-3671

REFERENCES


SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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