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RETROGRADE THORACIC DUCT ACCESS FOR EMBOLIZATION OF LYMPHATIC MALFORMATIONS IN A CHILD WITH CONGENITAL HEART DISEASE AND A PLEXIFORM THORACIC DUCT VARIANT

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ABSTRACT

The physiologic sequelae of the atypical vasculature in patients with congenital heart disease can result in potentially fatal lymphatic complications, especially after corrective cardiac surgery. Transcatheter embolization of the thoracic duct or lymphatic collaterals can reduce morbidity and mortality in these patients. While typically performed transabdominally via an antegrade approach, retrograde embolization may be preferable in cases where this is not feasible, including in rare variants of thoracic duct anatomy. We present a case of a child with severe chylothorax after congenital cardiac surgery who was found to have thoracic lymphatic malformations and a plexiform thoracic duct variant who underwent successful embolization of the malformations.

Keywords: Congenital heart disease; lymphatic malformation; embolization; aortic coarctation

Lymphatic complications in patients with congenital heart disease (CHD) are a cause of morbidity and mortality. Due to abnormal vascular anatomy, surgical procedures near the thoracic duct (TD), and elevated systemic venous pressure, many patients are at risk for lymphatic complications such as chylothorax, protein losing enteropathy, and plastic bronchitis (1). The morbidity and mortality from post-operative chylothorax after congenital cardiac surgery continues to be high (2).

TD embolization in these patients has been shown to be effective with symptomatic improvement (3). Typically, percutaneous embolization is performed transabdominally with catheterization of the cisterna chyli and then antegrade into the TD (1,4), however it is not always possible due to factors such as ability to visualize the cisterna chyli, the abdominal vasculature, and TD anatomy (4,5). In particular, the plexiform TD variant is technically difficult to advance wires and catheters from an antegrade approach, and in these cases, embolization must be performed via a retrograde approach (5). In addition to anatomic variants of the TD, previous surgery may preclude typical access routes to the cisterna chyli, creating the need for alternative approaches (6). We present an infant who developed severe, life threatening chylothorax after congenital cardiac surgery who underwent transcatheter retrograde cannulation of a plexiform TD and embolization of thoracic lymphatic malformations. The patient's family

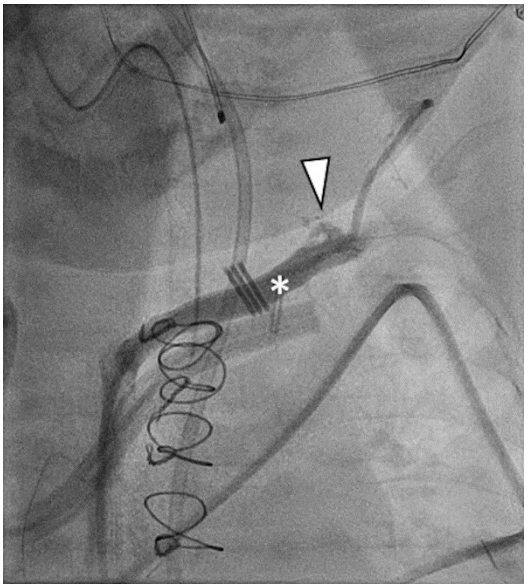

Fig. 1. *Angiogram of the innominate vein (*) with contrast refluxing into the proximal end of the thoracic duct and filling it retrograde (arrowhead).*

provided consent for this report.

CASE REPORT

The patient is a 6-month-old female born at 37 weeks gestation with aortic coarctation, partial atrioventricular septal defect, and mitral regurgitation. She was initially palliated with aortic angioplasty at 2 months old to defer definitive surgical intervention until at least 4 months old for more durable mitral valve tissue. She underwent successful surgical correction at 4 months old with repair of the aortic coarctation, closure of a primum atrial septal defect, and mitral valve cleft closure.

Post-operatively, there was a modest volume chylothorax which initially improved with a low-fat formula diet. However, there was an abrupt increase in chest tube output, and a large thrombus was identified in the superior vena cava (SVC) on echocardiography. She was brought to the catheterization lab and found to have multiple venous stenoses in the right iliac vein, inferior vena cava (IVC) below the renal veins, and SVC. The innominate vein pressure was 18 mmHg and the SVC pressure below the thrombus was 10 mmHg. She underwent angioplasty of all the venous stenoses with angiographic improvement and an immediate decrease in the innominate vein pressure to 12 mmHg. Stent placement was deferred due to her small size and anticipated resolution of the thrombi. Chest tube output decreased over the first 24 hours, but then increased to 200 mL/kg/day.

Due to the life-threatening nature of this volume of chylothorax output, she was brought back to the catheterization lab for attempted thoracic duct embolization. Due to her critical illness, presence of chest tubes, and temporary epicardial pacing leads, additional lymphatic imaging (lymphoscintigraphy, MRI) were not possible. In addition, positioning of chest tubes and pacing wires on her anterior abdomen, prevented transabdominal access to the cisterna chyli for antegrade cannulation of the TD. A transvenous, retrograde approach was planned.

At catheterization, lower body venous stenoses had not recurred, but SVC obstruction persisted and the innominate vein pressure was 17 mmHg. In anticipation of elevated innominate vein pressure contributing to reflux into the TD, no SVC intervention was undertaken yet. An angiogram in the innominate vein showed a widely patent vessel with reflux of contrast into the TD (*Fig. 1*). Using a JR 2.5 catheter, the proximal TD was engaged and a 45-degree SuperCrossΣ microcatheter (Teleflex, Inc, Wayne, PA) with a Runthrough® wire (Terumo Interventional Systems, Somerset, NJ) advanced into the TD (*Fig. 2A*). The microcatheter and wire were advanced around the arch of the TD and partway into the straighter part of the TD. The wire would not advance further toward the diaphragm and an angiogram showed that there was not a discrete central TD, but rather a plexiform TD (*Fig. 2B*). There was no obvious flow into the pleural space from the initial imaging. A WiggleΣ wire (Abbott, Inc, Plymouth, MN) was able to advance past the end of the catheter and enter the pleural space and the catheter easily followed. Angiography revealed markedly abnormal and dilated lymphatic channels, consistent with thoracic lymphatic

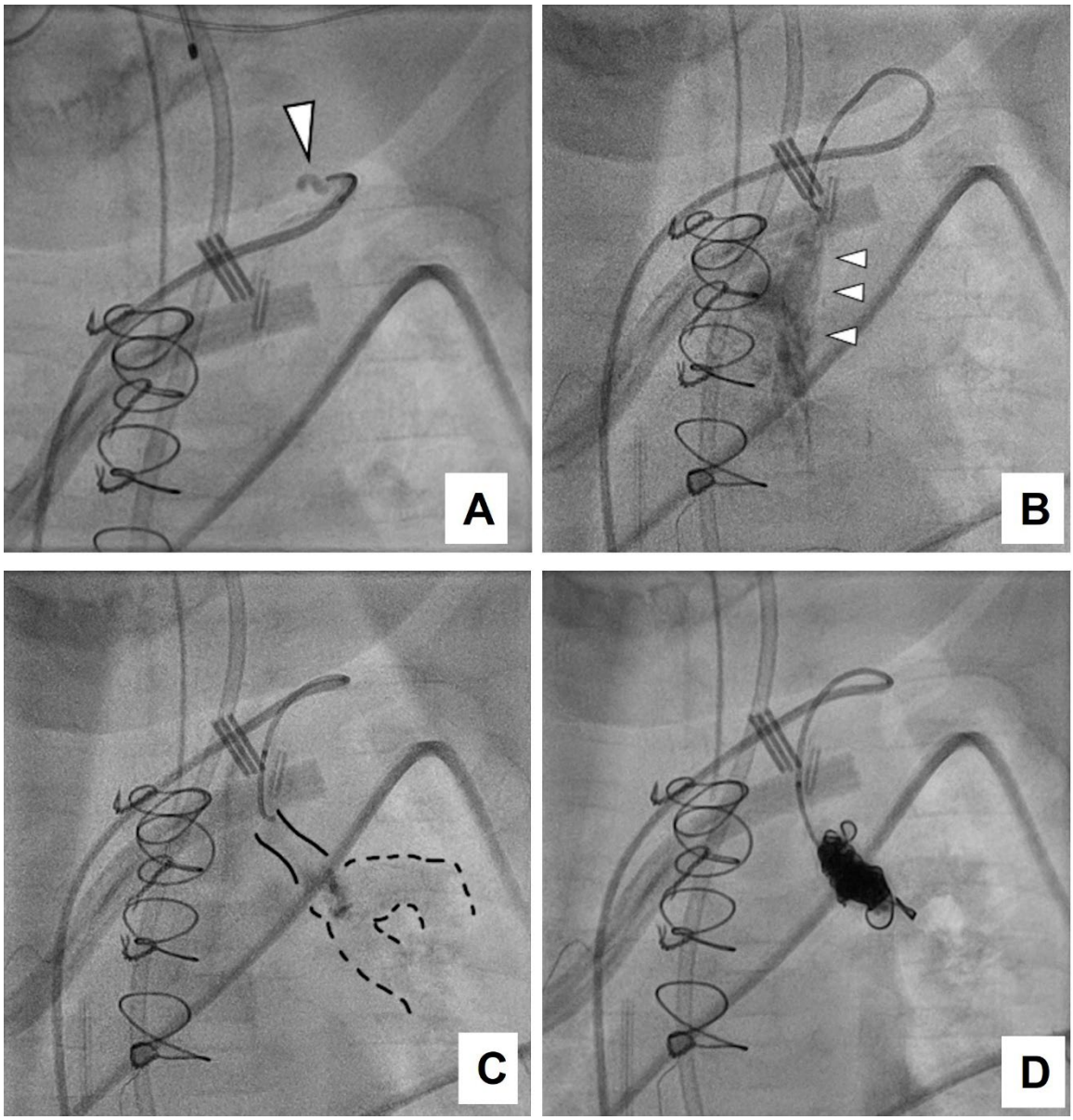

Fig. 2. *A) Retrograde cannulation of the thoracic duct with a microcatheter in the very proximal end (arrowhead) with an injection of contrast confirming the location. B) The microcatheter is more distal in the thoracic duct (arrowheads) which has a plexiform appearance. C) The microcatheter is now in a channel from the thoracic duct (solid lines) to large thoracic lymphatic malformations (dotted lines) within the left chest which communicate with the pleural space. D) After placement of embolization coils in the channel, there is near complete cessation of flow to the lymphatic malformations.*

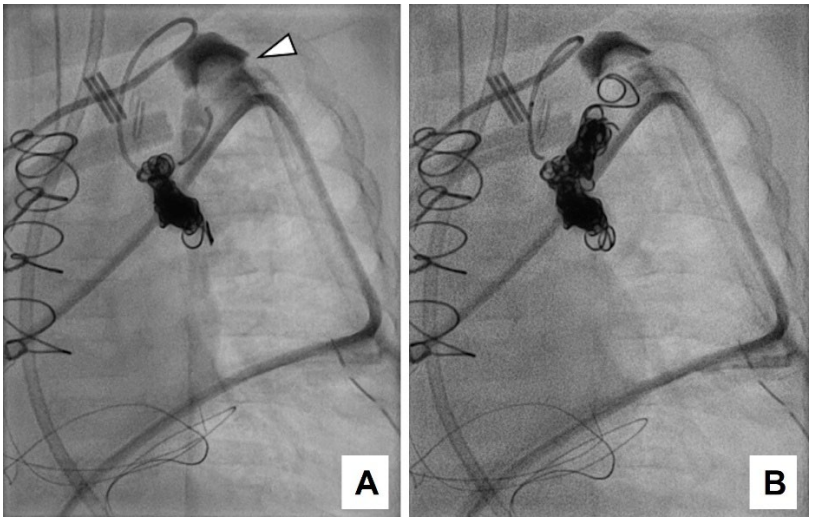

Fig. 3. *A) The microcatheter is in a second, superior channel from the thoracic duct to another thoracic lymphatic malformation (arrowhead) which communicates with the left pleural space. B) After placement of embolization coils in the superior channel, there is no further flow to the lymphatic malformation. Note, there is still some retained contrast in the pleural space from earlier angiograms.*

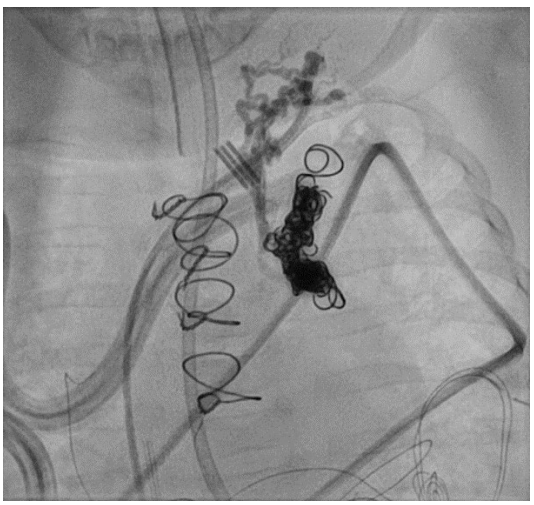

Fig. 4. *On follow-up catheterization, retrograde engagement of the thoracic duct and contrast injection shows normal flow and no communication with the left pleural space.*

malformations, which drained into the pleural space (*Fig. 2C*).

The plan was to attempt to only embolize these malformations, leaving the central TD intact to minimize the risk of chylous ascites. It was not anticipated that lymphatic fluid would clot in the fashion that blood typically does so a very dense coil pack of six AZUR™ CX coils (Terumo) was created to stop the abnormal lymphatic flow through the lymphatic malformation with reduction in flow to the pleural space (*Fig. 2D*). However, angiography now identified a superior malformation which also drained to the pleural space, and this was embolized with two AZUR™ CX coils with essentially complete cessation of flow (*Fig. 3*). No additional lymphatic collaterals were identified. SVC angioplasty was performed and the pressure decreased to 12 mmHg. Stenting was again deferred due to her small size.

There was improvement in the volume of chylothorax, however two days later she developed acute SVC syndrome. She was urgently brought back to the catheterization lab for intervention. There was persistent SVC thrombus and the innominate vein pressure was 16 mmHg. The TD was engaged retrograde in anticipation of additional embolization, however no further drainage into the pleural space was identified (*Fig. 4*). Given the recurrence of SVC stenosis after angioplasty alone, a 7-mm Express™ LD stent (Boston Scientific, Marlborough, MA) was placed in the SVC with

complete resolution of the stenosis and acute resolution of SVC syndrome. She was maintained on a low-fat formula diet and the chylothorax markedly decreased. She was eventually extubated and discharged home and is doing well at follow-up with no recurrence of pleural effusion or SVC syndrome. SVC stent redilation is planned as she grows.

DISCUSSION

This case highlights the potential for lymphatic malformations in children with CHD which may not be identified until after corrective cardiac surgery. Selective embolization of the thoracic lymphatic malformations prevented further communication to the pleural space and stopped the intractable, life threatening chylothorax.

While typically an antegrade approach for TD intervention is employed, complex, abnormal anatomy may preclude this and a retrograde approach may need to be considered. An advantage of an antegrade approach to TD intervention is the more direct route into the TD via the cisterna chylii with fewer turns to navigate a catheter through. In addition, since an antegrade approach "goes with the flow" there is less difficulty in advancing past one-way valves within the TD. A major disadvantage of an antegrade approach is the unpredictability of the size and location of the cisterna chylii and need for advanced imaging to identify it for percutaneous access. In critically-ill patients such as the one presented, advanced imaging may not be safe or even possible if there are implants that prevent MR imaging. There is also a risk of injury to the intestines and abdominal aorta during attempted percutaneous access of the cisterna chylii. With a retrograde approach, advanced imaging may not be needed in all cases and there is avoidance of abdominal structures, potentially minimizing the associated complications. This does come at the cost of potentially more complicated catheter routes and the need to advance catheters retrograde against closed one-way valves in the TD. There are clearly advantages and disadvantages for either approach, but having knowledge of both methods to access the TD will increase the chances of successful interventions.

CONFLICT OF INTEREST AND DISCLOSURE

All authors declare no competing financial interests exist.

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