Chapter 55
Surgical Management

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Introduction

This chapter focuses on surgical interventions for treating chyle reflux. As with many areas of vascular medicine, multidisciplinary management is the norm, with surgeons, medical specialists, and interventional radiologists all playing important roles. An understanding of chyle transportation from the bowel lacteals to the thoracic duct and venous system is important in planning treatment. When flow is disrupted, the anatomic location of a blockage or rupture in this ductal system may translate into chylous ascites, chylothorax, chylous cyst, chylopericardium, chyloptysis, chyluria, and skin lesions with chylorrhea. The goal of therapy is to restore balance to the lymphatic system through decreased lymph production, facilitation of physiological lymphatic pathways, or removal of regions of lymphatic dysplasia.

Drainage Procedures

Removal of pooled chylous fluid is another adjunct to surgical treatment. For example, compression garments and mechanical massage have shown good results in limiting and/or controlling lymphedema, cellulitis, and lymphangitis in extremities. When disrupted flow results in a chylothorax or chylous ascites, thoracentesis and paracentesis may be used to relieve symptoms of respiratory failure and improve hemodynamics. Wheeler et al. have described thoracentesis as a means of treating life-threatening neonatal tension chylothorax. To obtain continuous drainage, tube thoracostomy has been shown to be effective. In a study of 29 patients, Marts et al.
were able to combine medium chain triglyceride (MCT) therapy with chest tube placement for a cure rate of 79% of adult patients presenting with a traumatic or iatrogenic chylothorax. Though there is no consensus as to when drainage procedures should be abandoned in favor of surgery, most authors recommend surgery for persistent disease after two attempts at drainage.\textsuperscript{1-3}

The literature does not support drainage procedures as primary treatment for cutaneous chyle reflux, especially in the setting of lymphangiomyomatosis.\textsuperscript{4,5} Converting an internally draining system into an externally draining system may worsen chyle losses and hasten malnutrition.

**Image-Guided Approaches**

Dilated lymphatic channels and resultant chylorrhea are often the most symptomatic manifestation of chyle reflux. The cutaneous manifestation presents as milia-like lesions often in the external genitalia, thigh, or trunk.\textsuperscript{6,7} To address megalympathies, dilated channels, and “lakes,” luminal obliteration has become increasingly utilized. Toxic agents may be directly injected into the lumen of such vessels, causing obliterative lymphangitis. Commonly employed agents include tetracycline, doxycycline, alcohol, OK-432, and bleomycin.\textsuperscript{8,9} Alternatively, vessels may be obliterated by placing intraluminal coils. Two teams have advocated percutaneous lymphangiography-guided cannulation and embolization as primary treatment for chylous leak of the thoracic duct after head and neck surgery.\textsuperscript{10,11} Superficial lesions with lower degrees of lymphatic dilation have been successfully managed with the use of radiation therapy and lasers such as the carbon dioxide, yttrium aluminum garnet, and potassium titanyl phosphate laser. Carati et al.\textsuperscript{12} summarized their experience in a double-blinded randomized control trial with a low-level 904-nm laser in which one third of postmastectomy lymphedema patients demonstrated decreased limb volume and limb hardness.

**Open Surgical Approaches**

*Treatment of Cutaneous Chylorrhea and Chylorrhagia*

Removing a region of lymphatic dysplasia is often the most expedient route to symptomatic relief. This principle is true in almost all regions of the body, including the skin, bowel, thorax, and abdomen.\textsuperscript{13} Ruptured lymphatics are identified after the patient ingests a fatty meal and the leaking lymphatic ducts are ligated, oversewn, clipped, or excised with diathermy. Cutaneous lesions in the groin area have a high rate of recurrence, and multimodal therapy with excision and laser or sclerotherapy works well (Fig. 55.1).\textsuperscript{14} Percutaneous drainage tubes are often employed in the
postoperative phase to decrease the incidence of seroma formation. As previously stated, such drains transform an internally draining system into an externally draining system, and the amount of drainage must be monitored closely to avoid complications associated with high volume chyle loss.

A second surgical option is to reconstruct damaged lymphatics in an attempt to restore “physiological” lymphatic flow. The goal of physiological surgical intervention
is to bypass disrupted or malformed lymphatics and to restore physiological balance to the lymphatic system. Prior to undergoing physiological intervention, Kim et al.\textsuperscript{13} recommend that patients first undergo a trial of complex physical therapy (CPT) that includes manual lymph drainage, compression bandages, stockings or serial compression devices. Used primarily for its high rate of symptom amelioration, CPT also delays progression of chyle reflux to chronic interstitial edema, which is not amenable to reconstruction.\textsuperscript{15} However, Campisi\textsuperscript{16} believes that compression does not affect the microcirculation and attributes high nonresponse rates of 30–40% to this failure. Consequently, microsurgical techniques have been developed to create super-microsurgical anastomoses between lymphatics and venules.

The most common surgical approaches are through lymphaticovenous anastomoses and lymphatico-veno-lymphatic anastomoses. Both can be performed end-to-end or end-to-side, and both have been reported at vessel diameters of as little as 0.3 mm.\textsuperscript{17} In a report of 39 cases, Narushima\textsuperscript{18} found a positive correlation between the number of anastomoses to symptomatic improvement and decreased limb diameter. In another study, Kobayashi\textsuperscript{15} described a technique in which he telescoped, or invaginated, multiple small lymphatic channels into a single venule. Like Narushima, he documented both symptomatic and functional improvement in his cohort. The downside to microsurgical techniques is their technical difficulty and variable long-term patency rates.\textsuperscript{18}

**Treatment of Chylothorax**

With the development of a chylothorax, chest tube drainage is often implemented first to alleviate respiratory dysfunction. MCT diet, thoracentesis, and percutaneous obliteration follow as definitive therapy and/or helpful adjuncts. Videoscopic-assisted thoracoscopy (VATS) is a safe and effective technique to evaluate the entire thorax and allow for intervention based on the amount and location of output of the chylous leak.\textsuperscript{19} When output is less than 500 mL/day, pleural abrasion and pleurodesis with talc or other agents has been shown to be effective.\textsuperscript{20} Fibrin glue can be used during the VATS procedure to help manage chylous leakage when no focal drainage point can be determined.\textsuperscript{21} With diffuse drainage, partial pleurectomy has also been demonstrated to resolve symptoms.\textsuperscript{22}

For high output chylous drainage or drainage exceeding 1 L/day, ligation of the thoracic duct can be achieved with VATS or thoracotomy. The technique may involve the injection of methylene blue dye or consumption of a high fat content material, such as a butter/cream mixture, and observation for focal output. If diffuse drainage is seen, en masse tissue ligation can be undertaken as the thoracic duct generally follows posterior to the esophagus, and anteromedial to the azygos vein in the lower chest.\textsuperscript{23} Studies have shown a 90% resolution of the leak and earlier chest tube removal when the thoracic duct is ligated in both pediatric and adult populations.\textsuperscript{24,25} Alternatively, cannulation of the cysterna chyli with coil embolization of the thoracic duct may be performed. Partial or full response has been shown to be around 95% when either the cysterna chyli or thoracic duct can be identified and accessed.\textsuperscript{19}
Congenital chylothorax secondary to lymphatic aplasia, hypoplasia, fibrosis, and superior vena caval obstruction remains a clinical challenge. Serial thoracentesis was the classic treatment, but increased risk of infection, patient discomfort and the psychological effects of repeated trauma in children led to the development of new shunting procedures. The first clinical pleuropertitoneal shunts were reported in 1983 with a low-pressure ventriculoperitoneal shunt catheter system in five ventilator-dependent babies. Resolution of pulmonary failure and chylothorax was evident in four of the five patients. The Denver pleuropertitoneal shunt, which requires the patient to manually pump the fluid with a subcutaneous pump, has also been used for difficult cases of chylothorax. Side effects to these shunts include skin erosion, infection, and, most commonly, occlusion. In 1999, Wolff et al. described a modification of the pleuropertitoneal shunt with externalization of the pumping chamber that increased efficacy and comfort in utilizing this approach.

Retrograde chyle flow resulting from thoracic duct obstruction may be managed by a thoracic duct to azygos vein anastomosis. The procedure involves the lower thoracic duct (usually around 2–3 mm) and an end-to-end anastomosis with 8-0 or 10-0 non-absorbable suture.

Treatment of Chyloous Ascites

As with other disorders of chyle reflux, multimodal treatment is the rule with chyloous ascites. Percutaneous procedures, aimed at embolizing retroperitoneal lymphatics with needle disruption, are a first-line therapy in conjunction with an MCT diet. Cope et al. have shown a response in up to 42 (73.8%) of the study patients with percutaneous techniques. Patients with symptomatic chyloous ascites can be initially managed through serial paracentesis. However, if the drainage persists, a peritoneal–venous shunt can be utilized. The “LeVeen Shunt,” developed in 1974 to combat cirrhotic ascites, consists of a one-way pressure valve that shunts fluid from the peritoneal cavity to the internal or external jugular vein. However, complications in a nutritionally sub-optimal patient include high rates of wound infection, peritonitis, asymptomatic coagulopathy, disseminated intravascular coagulation (DIC), subclavian vein thrombosis, and electrolyte disturbances.

Adjuncts to Surgical Management of Recalcitrant Chyle Reflux

Octreotide, a somatostatin analog, has been shown in multiple studies to slow chylothorax output and hasten chylous fistula closure. The mechanism of action is thought to be a result of mild vasoconstriction of splanchnic vessels and the reduction in gastrointestinal secretions. The treatment can be delivered intravenously or subcutaneously. Side effects of this therapy include hypothyroidism, necrotizing enterocolitis, renal impairment, hyperglycemia, cramps, loose stools, and liver dysfunction.
Unpublished cases have shown promising results through the use of rapamycin in the inhibition of lymphangiomatosis. Rapamycin is a lipophylic macrolide antibiotic that inhibits mTOR, a serine/threonine kinase involved in VEGF-A expression and angiogenesis. Primarily investigated for its role in preventing tumor cell metastasis within the lymphatic system, rapamycin has been used to treat life-threatening lymphatic malformations at our institution (Fig. 55.2).
Summary

Treatment of chyle reflux, like most areas of vascular medicine, is in rapid evolution. Multidisciplinary management is the norm, with surgeons, medical specialists, and interventional radiologists all playing important roles.

References