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Intra-abdominal Chylovenous Bypass Treats Retroperitoneal Lymphangiomatosis

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Abstract

BACKGROUND

Retroperitoneal lymphangiomatosis (RL) is a rare form of primary lymphedema without existing treatment featuring aberrant retroperitoneal lymphatic proliferation. It causes recurrent cellulitis, repeated interventions, and poor life quality.

METHODS

Between 2012 to 2018, 44 primary lower-extremity lymphedema cases received lymphoscintigraphy, magnetic resonance imaging, and single-photon electron computed tomography to detect RL. RL patients underwent vascularized lymph node transfer (VLNT) for lymphedema and side-to-end anastomosis of lymphangiomatosis tissue to recipient veins as intra-abdominal chylovenous bypass (CVB) for chylous ascites. Complications, CVB patency, and quality of life were evaluated postoperatively.

RESULTS

Six RL patients (mean age 30.3 years) had chylous ascites and five had lower-extremity lymphedema. All CVBs remained patent, though one required re-anastomosis. All lymphedema cases underwent VLNT with 100% flap survival. Patients reported improved quality of life (p=0.023), decreased cellulitis incidence (p=0.041), and improved mean lymphedema circumference (p=0.043). All resumed normal diet and activity.

CONCLUSIONS

Intra-abdominal CVB with VLNT effectively treats retroperitoneal lymphangiomatosis with chylous ascites and extremity lymphedema.

DISCUSSION

CVB with VLNT physiologically returns extravasating lymph to intravascular space, preventing sequelae of displaced lymph. Screening primary lower-extremity lymphedema patients with MRI and SPECT could reveal a higher prevalence of RL than suspected and guide treatment of refractory lymphedema.

Keywords: retroperitoneal lymphangiomatosis, extremity lymphedema, chylovenous bypass, vascularized lymph node transfer

INTRODUCTION

Lymphangiomatosis is a rare condition of unknown incidence with diffuse involvement of all tissue types through hyperproliferation of lymphatic vasculature, causing multiple lymphangiomas. [1,2] This condition affects more children than adults. [2,3] Lymphangiomatosis is classified as either generalized lymphangiomatosis or Gorham Stout syndrome, with the former primarily involving soft tissue and viscera with secondary osseous involvement, while the latter features lymphovascular proliferation within bones with occasional soft tissue/viscera involvement. [3,4] Major complications of lymphangiomatosis include chylothorax, chylous pericardial effusions, and chylous ascites. [5,6] Chylous ascites has a poor prognosis without hope of spontaneous resolution. Persistent loss of chyle can also

cause nutritional imbalances. [7,8] Retroperitoneal lymphangiomatosis with concomitant lower-limb lymphedema has also been reported. [9] Current standards of care for these complications largely involve symptomatic relief by thoracentesis, pleurodesis, or paracentesis and combined therapy for any lymphedema. [10]

While medical and surgical management of retroperitoneal lymphangiomatosis exist, not all are effective or even resolve secondary chylous ascites or lower-extremity lymphedema. There is limited evidence for proposed medical treatments such as bisphosphonate, sirolimus, interferon, and radiotherapy. [2,7,8] There is also a paucity in interventional methods as well, especially for chylous ascites.

Kinmonth first described a series of patients with lower-extremity lymphedema caused by ectatic lymphatic channels in the abdomen, involving the retroperitoneum to the mesentery/intestines. Treatment included repairing leaks, ligating incompetent channels, and resection (e.g., intestinal resection,

hysterectomy). [9] Sclerotherapy has also been proposed with varied reception. [8, 10-12] Noel et al. reported 4 chylovenous anastomoses out of 26 surgical procedures in 30 patients with primary chylous disorders. [8] Campisi et al. opted for exeresis, CO_2 laser treatments, and chylovenous/lymphovenous shunts in 12 patients with primary chylous ascites (not all had retroperitoneal lymphangiomatosis). [9] Their shunt techniques varied, with direct lymphatic duct repair to subcutaneous venous anastomosis, chylous channel/cyst-to-venous shunt anastomosis, or lymphatic-to-venous anastomosis using vein grafts. Yet, anastomoses details were not reported, such as the level a technique was performed at or specific technique outcomes. [8,9]

To address such issues, our study aimed to investigate standardized diagnostic criteria and surgical outcomes of intraabdominal chylovenous bypass (CVB) with vascularized lymph node transfer (VLNT) for

retroperitoneal lymphangiomatosis with chylous ascites and concomitant lower-extremity lymphedema.

MATERIALS AND METHODS

Between 2012 and 2018, patients who presented with primary lymphedema or chylous discharge were retrospectively reviewed with institutional review board approval (IRB: 201801591B0) at Linkou Chang Gung Memorial Hospital. Each patient diagnosed with chylous ascites underwent assessment to discover the etiology, ranging from clinical measurements to imaging to detection of albumin levels. Ultrasound Doppler and computed tomography angiography (CTA) were utilized to rule out any proximal vascular lesions and to evaluate the patency of any VLNT for lower-extremity lymphedema. [13] Lymphoscintigraphy was performed, and Taiwan Lymphoscintigraphy Staging was used to interpret findings of partial or total lymphatic obstruction of the lower extremities. [14] Magnetic resonance imaging (MRI) was performed to confirm extravasation of contrast into the

peritoneal cavity and to visualize aberrant lymphatic tracts in the soft tissue. Since 2018, single-photon emission computed tomography (SPECT) was added to support the presence of abnormal retroperitoneal lymphatic tracts and evaluate disease extent. Indocyanine green (ICG) lymphography was performed in cases of mild extremity lymphedema.

When a patient was confirmed to have extremity lymphedema of Cheng's Grade II-IV, VLNT was suggested to first relieve the extremity lymphedema, followed by CVB to address the chylous ascites. The clinical rationale for performing VLNT instead of lymphovenous anastomosis (LVA) for extremity lymphedema was that prior studies had demonstrated that LVA is indicated for cases of Cheng Lymphedema Grade I-II lymphedema, while VLNT is indicated for Grade II-IV lymphedema. [14] Donor VLNT flaps were selected from the omentum or submental space.[15] Patients were monitored immediately after CVB in a microsurgical ICU ward for 3-5 days before being transferred to a regular ward. Patients were followed postoperatively and

evaluated for changes in limb circumference, chylous ascites, patency of the CVB by CTA, complications, body weight, and nutrition. Patients were also asked to rate their quality of life (QoL) using a visual analog score scaled 1 – 10 at their initial and latest clinical visit, with 1 being the poorest and 10 being the best.

TECHNIQUE FOR VLNT

The preoperative evaluation and technique of VLNT have previously been described by the authors.[16-18] Prior to the intra-abdominal CVB, the patency and effectiveness of any prior VLNT were tested by Doppler ultrasound and CTA. If the VLNT demonstrated impaired venous flow, it was revised, and the pedicle was confirmed to be patent with adequate flow at the time of intra-abdominal CVB.

TECHNIQUES FOR INTRAABDOMINAL CHYLOVENOUS BYPASS

The abdomens of patients were opened with a 15 cm incision for an exploratory laparotomy by a general surgeon (K-H Liu). Surgical exploration

then identified the potential site of damaged lymphatics responsible for chyle leakage in the retroperitoneal lymphangiomatosis soft tissue, as evidenced by focal milky fluid exudate. The precise site of leakage was confirmed under a microscope (Zeiss OPMI Pentero 800, Jena, Germany) at 20-25x magnification, with the location typically being between the level of the umbilicus and the anterior superior iliac spine around the ureter and internal iliac artery. A 4 mm diameter hole was created by bipolar cautery in lymphangiomatosis soft tissue as a controlled outlet for chyle. The ovarian vein was then identified in relation to the outlet. If the distance between the ovarian vein and outlet could not be bridged without tension, branches from the internal iliac vein were then identified as alternative recipient veins and dissected free. The designated recipient vein was separated distally. The distal end of the vein was then anastomosed to the artificial hole in a side-to-end fashion with 8-12 stitches using 9-0 nylon. Indocyanine green (0.1 mL) was injected into the distal site of retroperitoneal lymphangiomatosis. Fluorescent flow was seen under microscopy to travel from the

lymphangiomas through the anastomosis and to the recipient vein, verifying intraoperative CVB patency.

STATISTICAL ANALYSIS

Data before and after VLNT and CVB, including data on clinical monitoring of chylous ascites, body weight, subjective QoL measures, and circumferential sizes of limbs, were collected and analyzed with IBM SPSS Statistics 20 statistical software (IBM Corporation, Armonk, NY). Data were reported as the mean ± standard deviation, and the circumference of the lower limbs measured 15 cm proximal to the patella, 15 cm distal to the patella, and 10 cm proximal to the ankle. Comparisons between groups were performed using the Wilcoxon signed rank test. A two-tailed p-value < 0.05 was considered statistically significant.

RESULTS

A total of 44 primary lymphedema patients were evaluated at Linkou Chang Gung Memorial Hospital between 2012 and 2018. 6 of the 44 patients

were diagnosed with retroperitoneal lymphangiomatosis (incidence: 13.6%). There were 7 primary lymphedema patients who received abdominal SPECT and MRI with results possibly indicative of retroperitoneal lymphangiomatosis, prompting exploratory laparotomy. Exploration found retroperitoneal lymphangiomatosis in 6 of the 7 patients. The specificity of SPECT plus MRI was 85.7% (6/7) in determining retroperitoneal lymphangiomatosis in primary lymphedema patients.

The average age of patients was 30.3 years (range 14- 55 years) (Table 1). All were female. Four patients presented with unilateral lower-extremity lymphedema, with 2 patients presenting with chyle leakage in the thigh, and one presenting with bilateral lower-extremity lymphedema (Fig. 1, Table 1). The mean preoperative Cheng Lymphedema Grade of the six lymphedematous limbs was 2.7 (range 1 - 4). Of the six limbs, four underwent VLNT prior to CVB, one underwent CVB only, and one underwent VLNT simultaneously with the CVB (Fig. 1). The average time between CVB and

VLNT was 23.6 ± 23.9 months (Table 1). Ultrasound Doppler studies showed patency and good flow in all pedicle anastomoses in the six VLNTs without stenosis, and lymphoscintigraphy demonstrated uncompromised VLNTs. All patient MRIs revealed chylous ascites, and lymphoscintigraphy confirmed partial or total obstruction in the lower extremity (Fig. 2).

The average albumin level changed from 3.1 ± 0.6 g/dL pre-CVB to 3.3 ± 0.7 g/dL post-CVB (p=0.500). The mean body weights pre- and post-CVB were 56.7±14.1 kg and 53.5±10.7 kg, respectively (p=0.07). Mean limb circumference after VLNT and CVB significantly improved by 4.2 ± 2.2 cm (p=0.043), and improved 2.6 ± 2.9 cm above the knee (p=0.068), 5.9 ± 4.7 cm below the knee (p=0.042), and 3.3 ± 2.2 cm above the ankle (p=0.078). Mean cellulitis incidence significantly declined from 1.9 ± 1.8 to 0.1 ± 0.4 times/year postoperatively (p=0.04) (Table 2).

Immediate patency of the CVB was verified in all patients through intraoperative ICG microscopy (Fig. 2). The average cohort length of stay

after CVB was 14.2±4.1 days (Table 3). Before discharge, all patients had <30 mL/day of chylous output from abdominal drains. No abdominal drains needed to be reinserted. One patient experienced post-CVB complications of perforation of the jejunum and thrombosis of the chylovenous anastomosis site, which required resection with primary repair and thrombectomy with repair of the jejunum and CVB, respectively. By discharge, all patients were able to return to normal activity. Patients were able to tolerate a regular diet instead of a low-fat diet by 6 months postoperatively without recurrence of chylous ascites. The resolution of chylous ascites in addition to patency of the CVB was verified through CTA at postoperative visits for all patients. Additionally, during follow-up surveillance, five of six patients were able to discontinue the use of compression garments for lymphedematous extremities by 6 months, with one patient continuing to require compression garments for 8-10 hours during the day for thigh lymphedema (Fig. 4) (Table 3). The mean QoL score significantly improved from 3.4 ± 0.8 at before the operation to 5.7 ± 1.3 after the operation (p=0.023) (Table 2).

DISCUSSION

Utilization of abdominal MRI was initiated due to the VLNT-refractory nature of extremity lymphedema with additional abdominal symptoms. MRI demarcated soft tissue changes and lymphatic proliferation of lymphangiomatosis. SPECT in the last four patients helped further elucidate the disease extent. Overall, lymphoscintigraphy, MRI, and SPECT were able to definitively demonstrate chylous ascites secondary to retroperitoneal lymphangiomatosis (Fig. 3).

Once VLNT was demonstrated to be inadequate for the concomitant lower-extremity lymphedema, CVB was performed as a potential physiologic treatment for the extravasating lymph causing the lymphedema and chylous ascites. The CVB procedure, unlike VLNT, appeared to be effective in treating the chylous ascites, as supported by the absence of clinically detectable ascites with imaging validation in all six patients. Additionally, following CVB, there was an additional significant reduction in leg circumference below the

knee beyond the VLNT-related reduction (p=0.042). The only exception to this trend was patient 5, whom removed her compression garments only during her pre-CVB measurements and therefore had an unknown true baseline. The single patient in our study unable to discontinue wearing compression garments postoperatively opted to do so for comfort, as her occupation requires her to stand for prolonged periods of time.

A potential mechanism for our observations is that the CVB technique creates a controlled outlet for high-volume chyle leakage from the lymphangiomatosis. This opening enables the drainage of chyle/lymph from the retroperitoneal space directly back to the venous system without passing into the thoracic ducts. Joining the lymphatic system with the systemic venous system thereby functions as a near-physiologic solution that not only prevents ascites but also limits protein loss and excessive lymphatic drainage to the extremities, which could otherwise contribute to hypoalbuminemia, lymphedema, infection, and soft tissue or bone damage.

Interestingly, although CVB theoretically enables the return of lost products to systemic circulation, not all patients presented with hypoalbuminemia pre-CVB, and not all patient presented an improvement in albumin level post-CVB ($p=0.500$). The lack of consistent hypoalbuminemia in the series could be a reflection of the duration of chylous ascites, with perhaps normal albumin levels reflecting earlier stages of retroperitoneal lymphangiomatosis. Persistently low albumin levels at follow-up may be due to slow replenishment after prolonged loss in the form of chylous ascites.

Patient body weights may decrease after VLNT but increase after CVB. It is difficult to determine the cause of a significant difference when the patients have undergone two procedures. However, the fact that all but one patient could tolerate a normal diet without weight gain should be considered. The 4.6 kg gain in patient 1 is likely actually a reflection of clinical improvement, as she was underweight at 33.4 kg as a fully grown adult as a result of her

severe, chronic malnourishment from chylous leakage alongside her numerous other disease states.

Although the time between when VLNT and CVB were performed varied, the rate of resolution of the chylous ascites or lower-extremity lymphedema improvement remained unchanged. These results do not indicate that VLNT was unhelpful, however; patients still had decreased chyle output and decreased lower extremity circumference with VLNT alone. Further studies with multivariate component analysis would be better suited to parse the contributions of VLNT vs CVB.

One concern that was later considered by the authors during the course of the series was that the omental lymph node transfer may not necessarily be effective for the extremity lymphedema, as the omental lymph nodes mainly drain the abdominal chyle. If the lower-extremity lymphedema observed was truly due to chylous ascites causing lymphatic leakage into the legs, then the omental VLNT would be ineffective, as drainage of chyle in situ proved

inadequate. In patients 4 and 5, the lower extremities underwent lymph node transfer without removing nodes from the abdomen. Patient 2 did not have lower-extremity lymphedema and thus did not undergo VLNT, although it is possible she could have eventually developed lymphedema had her disease not been caught early. The measurements of patient 1 could not be standardized given her above-knee amputation.

There are many challenges to performing a CVB: coordination with a general surgeon for exploration and reconstructive microsurgeon for anastomosis; a deep, chyle-flooded operative field; intestinal peristalsis causing movement; limited recipient vessels; requirement for longer microsurgical instruments; and having to perform anastomosis of a vein to a literal hole in the wall of the retroperitoneum under a microscope. The anastomosis itself is technically challenging, requiring multiple interrupted sutures while avoiding leakages and controlling internal bleeding. Continuous suturing is inadvisable, as this technique can narrow the anastomosis,

resulting in low flow or thrombosis that could lead to inadequate chyle drainage.

Maegawa et al. described using vein grafts to resolve cutaneous chyle leakages in the lower limb from the dilated lymphatic duct to the adjacent saphenous vein at two levels. [19] In comparison, we identified the intraabdominal abnormalities as the source of chylous ascites and chose to perform CVB from the incompetent tissue deep in the abdominal cavity to a low resistance path into the local venous system. By doing so, we bypassed the chyle leak and prevented continued lymphatic output from the lymphangiomas, resulting in complete treatment.

This study also provides a descriptive framework of how one might diagnose retroperitoneal lymphangiomatosis, particularly if there is no overt primary complaint of chylous ascites (Fig. 3). Further studies of the outcomes of CVB on retroperitoneal lymphangiomatosis with chylous ascites and lower-

extremity lymphedema are needed to validate the operative safety, outcomes, and long-term resolution.

CONCLUSIONS

Retroperitoneal lymphangiomatosis with chyle leakage and lower-limb lymphedema is a disease that is difficult to diagnose and treat. Lymphoscintigraphy, SPECT, and MRI are the recommended imaging modalities for diagnosing retroperitoneal lymphangiomatosis in primary lymphedema patients and/or patients with chylous ascites. Although the data are limited, CVB with vascularized lymph node flap transfer is recommended as a first-line measure for effective shunting of chyle into the intraabdominal venous system, simultaneously resolving the secondary lower-extremity lymphedema.

Synopsis

Shunting chyle from an artificial chyle outlet made in retroperitoneal lymphangiomatosis tissue into the venous system by venous anastomosis intra-abdominally resolves the incidence of chylous ascites and treats concomitant lower extremity lymphedema with lasting results with low rates of complications.

Data Availability

Raw data were generated at Chang Gung Memorial Hospital. Derived data supporting the findings of this study are available from the corresponding author on request.

Conflict of Interest

None

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Figure 1. Case 1: Initial Imaging and Follow-up Imaging. A) Pelvic X-ray revealed left femoral shaft and head fractures with an internal prosthesis and wiring fixation of a left iliac osteolytic fracture. B) MRI showed extravasation of contrast media to the left pelvis, left femoral area, and bilateral paraspinal regions, consistent with retroperitoneal lymphangiomatosis. The soft tissue of the left pelvis and thigh showed characteristic honeycomb lymphedematous fibrosis. C) and D) Computed tomography angiography revealed a patent chylovenous bypass (indicated by the yellow arrow) at the left iliac region without chylous ascites at 3 weeks (C, coronal section) and 3 months (D, sagittal section) postoperatively.

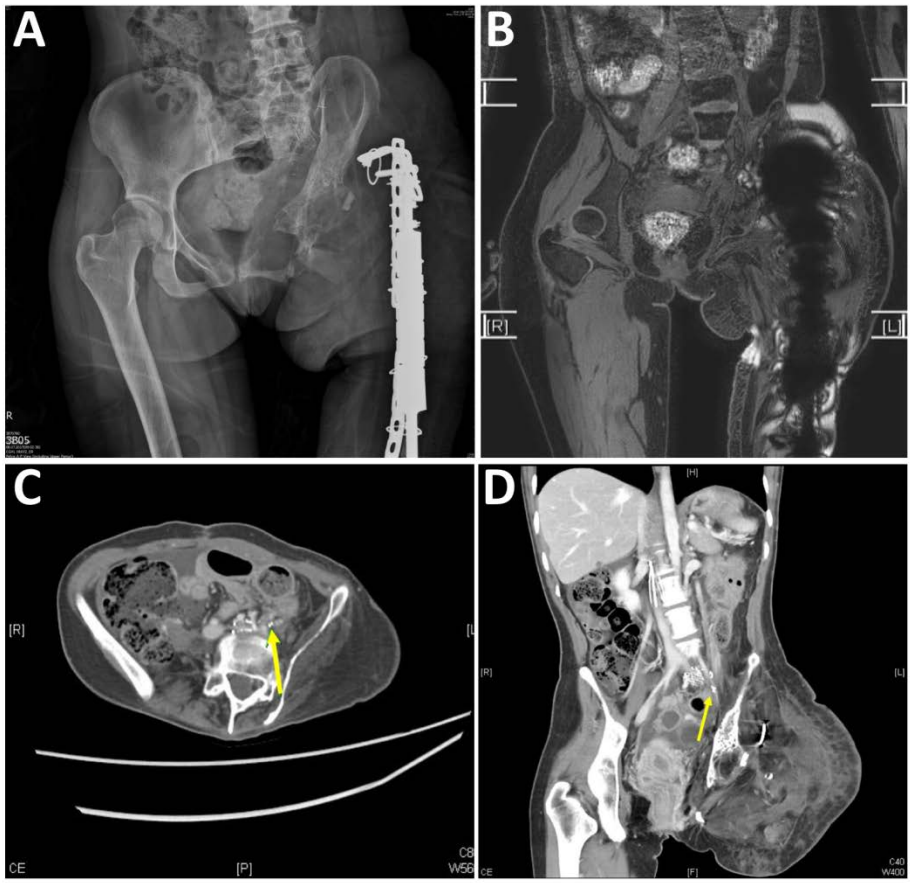

Figure 2. Case 5: Lymphoscintigraphy, Single-Photon Emission Computed Tomography, and Intraoperative Images with Indocyanine Green Confirmation. A) Anterior and posterior planar imaging of the lower extremities at 5 minutes and 2 hours post subcutaneous injection of 0.5/0.5 mCi Tc-99m phytate. Lymphoscintigraphy revealed reduced inguinal/pelvic lymph node uptake and dermal backflow over the entire left lower limb with no major abnormalities in the right lower limb in comparison. B) Accumulated tracer along the left iliac vessels/paraaortic area was suggestive of possible lymphangiomatosis with dilated lymphatic ducts in single-photon emission computed tomography. C) Chyle (white arrow) was actively leaking from the soft tissue of the retroperitoneal lymphangiomatosis intraoperatively at the prespinal area. D) The chylovenous bypasses were performed with 9-0 Nylon in a side-to-end fashion (yellow arrows indicating side-to-end anastomosis sites). E) Intraoperative ICG imaging confirmed patency of the chylovenous bypasses.

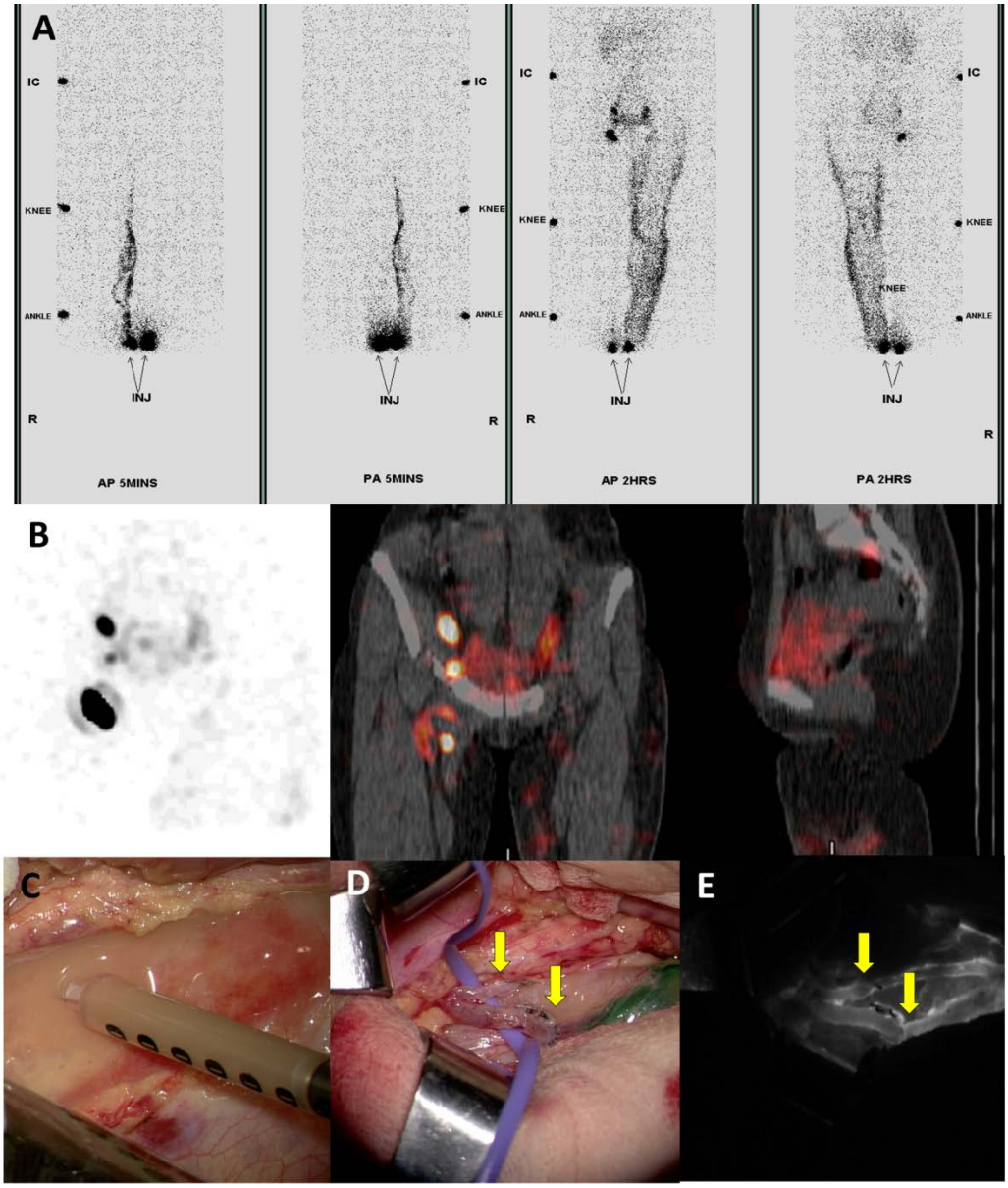

Figure 3. Proposed Algorithm for the Diagnosis and Treatment of Retroperitoneal Lymphangiomatosis Stepwise diagnostic pathway for work-up, differential diagnoses of possible causes of lower extremity lymphedema.

Figure 4. Pre-operative and Post-operative Photographs of the Left Lower Extremity Lymphedema of Patient 5 A) Pre-operative evaluation of patient 5's left lower extremity immediately after removal of her compression garments, demonstrating characteristic fibro-adipose tissue deposits and lymphedematous enlargement of the left leg throughout from thigh to foot. B) Post-operative day 15 imaging of the patient following her vascularized lymph node transfer and chylovenous bypass, showing reduction in circumference around the region above her knee, below her knee, and over the dorsal aspect of her foot, with protuberance around the ankle largely due to the VLNT. C) Follow-up imaging of the patient's left leg at 11 months with measurements above the knee, below the knee, and above the ankle still slightly improved compared to pre-operative measurements despite not wearing compression garments since surgery and standing for long stretches constantly at her occupation.

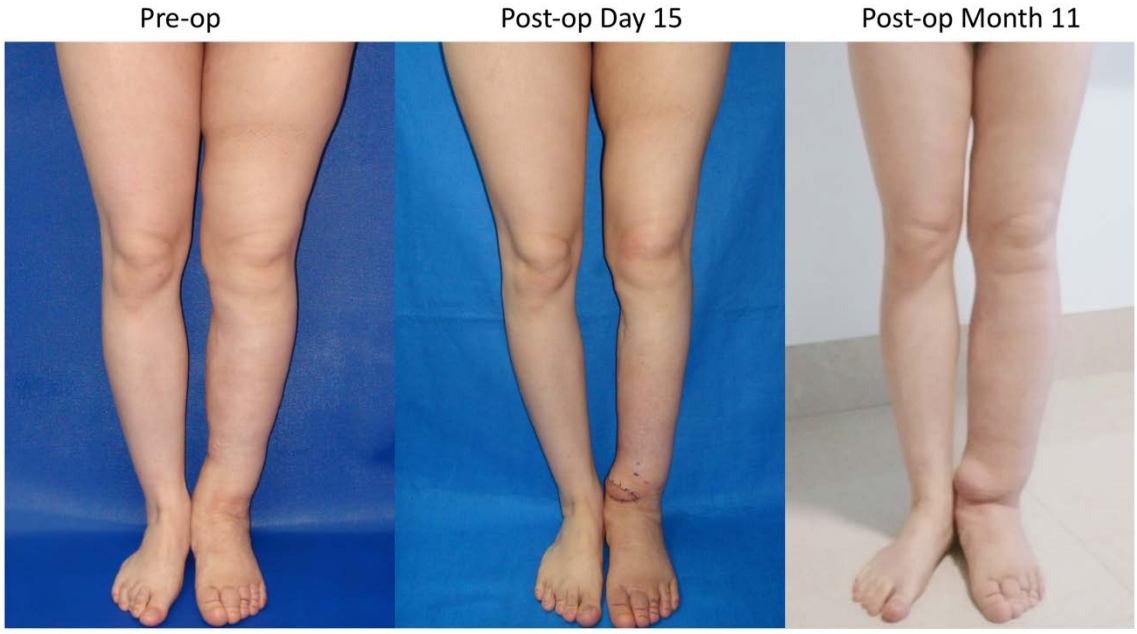

Table 1. Patient Demographics and Assessment Modalities for Retroperitoneal Lymphangiomatosis with Lower-Extremity Lymphedema

			Extremity Lymphedema				*Work-Up for Retroperitoneal Lymphangiomatosis*						
Case	**Age (Years)**	**Gender**	**Extremity Lymphedema**	**Cheng Lymphedema Grade**	**Partial /Total Occlusion on LSG (Grade)**	**VLNT**	**CTA / Doppler**	**Months from VLNT to CVB**	**Pre-op Albumin**	**SPECT**	**Pre-op MRI**	**Abdominal Findings**	**Follow-up Duration (Months)**
1	21	Female	L thigh	1	Total (T5)	Submental VLNT to lower L abdomen	Yes	63	Yes	No	Yes	Retroperitoneal lymphangiomatosis with lymphatic occlusion below the L common iliac chain	15
2	14	Female	None	-	N/A	No	Yes	-	Yes	No	Yes	Lymphangiomatosis involving the pancreas, spleen, duodenum, small bowel, and L colon	13
3	46	Female	L leg	1	Partial (P4)	Omental VLNT to R thigh	Yes	18	Yes	Yes	Yes	Lymphangiomatosis extending to the retroperitoneum	12
4	16	Fem al	R thigh	4	Total (T5)	Omental VLNT	Yes	12	Yes	Yes	Yes	Lymphangiomatosis extending	9

		e				to L ankle						to the retroperitoneum	
5	30	Female	L leg	2	Total (T5)	Submental VLNT to L ankle during CVB	Yes	0	Yes	Yes	Yes	Lymphangiomatosis extending to the retroperitoneum	11
6	55	Female	R leg L leg	R: 4 L: 4	R: Total (T5) L: Total (T5)	Submental VLNTs to L&R ankles, revision with CVB	Yes	25	Yes	Yes	Yes	Retroperitoneal and pelvic lymphangiomatosis with lymphatic outflow obstruction	4
Total/Mean	30.3 ± 16.8	-	-	2.7 ± 1.5	6	6	6	23.6 ± 23.9	6	4	6	-	10.7 ± 3.8

VLNT = vascularized lymph node transfer, CVB = chylovenous bypass, LSG = lymphoscintigraphy, MRI = magnetic resonance imaging, SPECT = single-photo emission computer tomography, CTA = computed tomography angiography, R = right, L = left

Table 2. Outcomes of Chylovenous Bypass and Vascularized Lymph Node Transfer on Retroperitoneal Lymphangiomatosis with Extremity Lymphedema

	Cellulitis Incidence (Times/Year)		*Body Weight (kg)*		*Albumin (g/dL)*		*Pre-CVB Limb Circumference (cm)*				*Post-CVB Limb Circumference (cm)*				*Improvement of Limb Circumference (cm)*			
Case	**Pre-CVB**	**Post-CVB**	**Pre-CVB**	**Post-CVB**	**Pre-CVB**	**Post-CVB**	**AK**	**BK**	**AA**	**Mean**	**AK**	**BK**	**AA**	**Mean**	**AK**	**BK**	**AA**	**Mean**
1	0	0	33.4	38	2.48	3.6	-	-	-	-	-	-	-	-	-	-	-	-
2	0	0	55	50	2.49	2.72	-	-	-	-	-	-	-	-	-	-	-	-
3	1	0	53.3	51	3.27	4.17	46	42.5	27	38.5	43.5	30.5	22.5	32.2	2.5	12.5	4.5	6.5
4	4	1	66.3	59.9	2.76	2.49	64.5	56	41.5	54	57	49	38	48	7.5	7	3.5	6
5	1	0	57	52	3.93	3.41	46	36	30	37.3	44	35.5	26.5	35.3	2	0.5	3.5	2
6	R: 4 L: 3	R: 0 L: 0	75.1	70	3.71	-	R: 42 L: 42	R: 55 L: 47.5	R: 47 L: 39	R: 48 L: 42.8	R: 42 L: 41	R: 48 L: 45	R: 40 L: 41	R: 43.3 L: 42.3	R: +0 L: +1	R: +7 L: +2.5	R: +7 L: -2	R: +4.7 L: -1.8
Me	1.9	0.1	56.	53.	3.1	3.3	48	47	36	44.	45	41	33	40.	2.6	5.	3.	4.2

an ± SD	± 1.8	± 0.4	7 ± 14.1	5 ± 10.7	± 0.6	± 0.7	.1 ± 9.4	.4 ± 8.5	.9 ± 8.3	1 ± 6.9	.5 ± 6.5	.6 ± 8.2	.6 ± 8.5	2 ± 6.4	± 2.9	9 ± 4.7	3 ± 3.2	± 2.2
P-values	Pre- vs Post-CVB Cellulitis Incidence *p* = 0.041	Pre- vs Post-CVB Body Weight *p* = 0.074	Pre- vs Post-CVB Albumin *p* = .500	Note: p-values for circumference changes calculated per limb														Pre- vs Post-CVB AK Circumference improvement: *p* = 0.068 Pre- vs Post-CVB BK Circumference improvement: *p* = 0.042 Pre- vs Post-CVB AA Circumference improvement: *p* = 0.078 Pre- vs Post-CVB Mean Circumference improvement: *p* =0.043

P-values determined using 2-tailed Wilcoxon signed rank tests. Dashes represent patients without data for that category. Measurements were missing in case 1 despite thigh lymphedema due to above-knee amputation of that limb. R = right, L = left, CVB = chylovenous bypass, VLNT = vascularized lymph node transfer, AK = above knee, BK = below knee, AA = above ankle. Case 6 was missing post-CVB albumin data. Body weight and limb circumference values were taken from the most recent follow-up.

Table 3. Surgical Outcomes of Chylovenous Bypass for Retroperitoneal Lymphangiomatosis with Chylous Ascites

Case	Operative Staging	Pre-CVB Evaluation	Intra-op CVB ICG Check	Days to Discharge Post-CVB	Post-Op Complications	Post-CVB Garment Usage	Normal Diet <6 Months Post-CVB?	Normal Activity by Discharge?	Post-CVB Evaluation	
									CTA	Clinical
1	VLNT then CVB	1. Milky discharge from abdomen, L thigh, L groin 2. Failure to thrive 3. L femur fracture	Patent	10	1. Thrombosis of CVB with thrombectomy 2. Perforated jejunum with resection, anastomosis	None	Yes	Yes	Patent L ovarian vein	1. Minimal ascites 2. VLNT uncompromised
2	CVB only	1. Protein-losing enteropathy 2. Recurrent diarrhea 3. Diffuse mesenteric heterogenous cystic lesions	Patent	18	None	None	Yes	Yes	Patent L ovarian vein	1. Minimal ascites
3	VLNT then CVB	1. Progressive R thigh swelling of 1 year with hyperpigmentation 2. Intermittent	Patent	13	None	Compression bandages 8-10 hours/day	Yes	Yes	Patent L, R ovarian veins	1. Reduced lymphangiomatosis on MRA POD10 2. Minimal ascites

		dull L upper abdominal pain with nausea								3. VLNT uncompromised
4	VLNT then CVB	1. Progressive L lower limb swelling of 2 years 2. Sensation of increasing abdominal fullness of 3 months	Patent	17	None	Compression bandages for 3 months then DC'ed	Yes	Yes	Patent internal iliac vein	1. Minimal ascites 2. VLNT uncompromised
5	VLNT and CVB	1. L lower limb swelling >20 years 2. L foot lymphovenous anastomosis in 2003, L leg debulking in 2008 without improvement	Patent	18	None	Compression bandages for 6 months then DC'ed	Yes	No	Patent L, R gonadal veins	1.Decreased lymphangiomatosis on CTA POD14 2. Minimal ascites 3. VLNT uncompromised
6	VLNT then CVB*	1. Bilateral lower limb swelling of >10 years 2. History of lower limb cellulitis	Patent	9	Congestion of VLNT with successful salvage	Compression bandages for 26 days then DC'ed	Yes	Yes	Patent L ovarian, R gonadal veins	1. Minimal ascites 2. VLNT uncompromised
Total/ Mean	-	-	6	14.2±4.1	3	-	6	5	-	-

Average hospital stay after CVB = 14.2±4.1 days. POD = Post-op Day. DC'ed = Discontinued. *Bilateral VLNT performed prior to CVB