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Intra-abdominal Chylovenous Bypass Treats Retroperitoneal Lymphangiomas

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OBJECTIVE: This study aimed to establish proper work-up and efficacious treatment of retroperitoneal lymphangiomas via intra-abdominal chylovenous bypass with vascularized lymph node transfer.

BACKGROUND: Retroperitoneal lymphangiomas is a rare form of primary lymphedema featuring aberrant retroperitoneal lymphatic proliferation. Currently, no protocols exist to attenuate its chylous discharge which causes recurrent cellulitis, repeated temporary interventions, and poor quality of life.

METHODS: Between 2012 and 2018, 44 primary lower-extremity lymphedema patients were diagnosed by lymphoscintigraphy. Magnetic resonance and single-photon electron computed tomography were used to detect retroperitoneal lymphangiomas. Patients with thigh chylous leakage or lower-extremity lymphedema underwent vascularized lymph node transfer (VLNT). Those with chylous ascites underwent intra-abdominal chylovenous bypass (CVB), anastomosing chyle-leaking tissue to recipient abdominal veins. Complications, CVB patency, and quality of life were evaluated postoperatively.

RESULTS: Six patients were diagnosed with retroperitoneal lymphangiomas (mean age 30.3 years, body weight 56.7 kg) presented with chylous ascites, and five presented with concomitant lower-extremity lymphedema. All six CVBs were patent, with one requiring reanastomosis. Four patients with unilateral and one with bilateral extremity lymphedema underwent VLNT with a 100% flap success rate. Follow-up

computed tomography angiography demonstrated patent chylovenous anastomoses. Patients reported improved quality of life ($p=0.023$), decreased cellulitis incidence (1.9 ± 1.8 ; $p=0.041$), and improved mean lymphedema circumference ($p=0.043$). All resumed normal diet and activity. To the best of our knowledge, this is the first report of CVB resolving chylous ascites from retroperitoneal lymphangiomas.

CONCLUSIONS: Intra-abdominal CVB with VLNT is an effective intervention for treating retroperitoneal lymphangiomas with chylous ascites and extremity lymphedema.