

A RATIONAL APPROACH TO THE MANAGEMENT OF LYMPHEDEMA

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ABSTRACT

Because of the complexity of peripheral lymphedema disorders and the variability in severity and clinical appearance, no standardized management plan of these patients is universally accepted. Based on our lengthy experience, we attempt in this report to outline a protocol for "staging" and managing lymphedema including the role of microsurgical lymphatic angioplastic shunts.

Because of complex etiologies and pathogenesis of peripheral lymphedema and variable clinical presentations (1), a proper standardization or "staging" of lymphedema to assess treatment and

clinical outcome is desirable. Based on our vast clinical experience with an ongoing registry (since 1973) we have proposed a classification of lymphedema based on four stages (Table 1).

Nonoperative therapy includes "non-specific" drugs depending on circumstances such as antibiotics, anti-inflammatories, diuretics (low dosage and short lived), emollients (anti-liposclerotics), and more "specific" agents such as connective tissue enzymes (e.g., mucinolytic derivatives) and "lymphotropes" such as benzopyrones and rutosides. Although these agents can be used at any stage, their beneficial value is most noticeable in the first three stages. Physical therapy and manual massage as advocated by Vodder, Földi, and

Table 1
Clinical Staging of Lymphedema and Therapeutic Options

STAGE	OPTION
I. Mild and intermittent edema	Conservative therapy*
II. Moderate and persistent lymphedema	Conservative therapy Microsurgical lymphatic shunts
III. Unremittent and intense edema	Conservative therapy Pneumatic compression (peristaltic pump) Microsurgical lymphatic shunts
IV. Fibrosclerotic lymphedema (elephantiasis)	Conservative therapy Intensive cyclic pressotherapy Microsurgical lymphatic shunts "Debulking" (reductive plastic surgery)

*Includes manual lymph drainage, external compression stockinettes, appropriate drugs

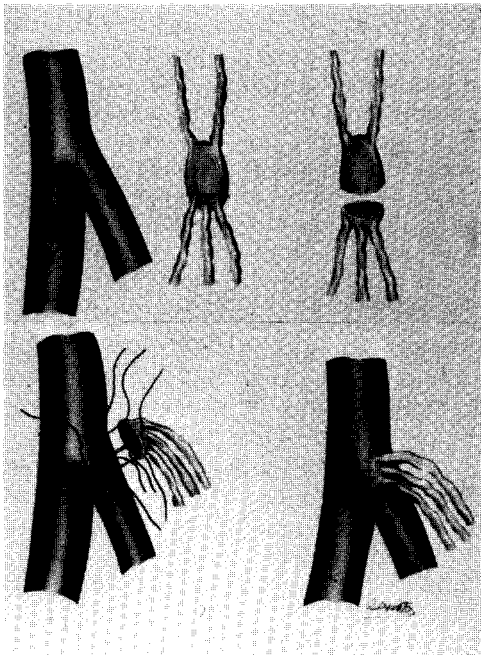


Fig. 1. Schematic drawing of lymph nodal capsular-venous anastomosis.

Leduc (2) has been in our experience most useful in the earlier stages or after operation. More intensive and prolonged mechanical and compression therapy with uniform, sequential "peristaltic" pressure pumps (3) has provided the most benefit in advanced stage lymphedema as well as after microsurgical (lymphatic-shunt) operations for advanced disease (4). Experience with thermotherapy (5) also has yielded improvement not only with lymphedema due to filariasis but also for postlymphangitis obliterans lymphedema. It is not entirely clear that heat enhances lymph flow as opposed to merely softening an otherwise indurated extremity. Microsurgical techniques to enhance the flow of stagnant lymph is reserved for



Fig. 2. Intraoperative photograph of end-to-side lymphatic-venous anastomosis (40x magnification) (left) and multiple end-to-side lymphatic-venous anastomoses (right).

patients with persistent lymphedema in whom nonoperative protocols have failed to improve the clinical situation. Direct "shunts" (6) include lymph nodal-venous and our modified lymphatic capsular-venous anastomoses (7) (Fig. 1) and, lymphatic-venous anastomoses (Fig. 2) especially end-to-side using Degni's needle (8). The latter approach has personally been modified using the outlet of a secondary branch of the recipient vein as an entry site for the lymphatic. In this way, the risk of stenosis at the lymphatic-venous entry site is minimized since phlebotomy in the main anastomosing vein and muscular fiber contraction is avoided.

Accurate angiologic assessment is critical especially with new and noninvasive techniques and particularly when microsurgical reconstructive operations are considered. Thus, lymphatic and venous isotope scintigraphy, Doppler venous flow metrics and manometry and occasional conventional (oil contrast) lymphography are indispensable to management especially in patients with unremitting or advanced disease. Moreover,

Table 2
Assessment of Clinical Outcome

Result	Edema Regression	Functional Improvement
Excellent	>75%	Complete recovery
Good	50-75%	Almost complete recovery
Fair	25-50%	Partial recovery
Poor	Unchanged or worse	None

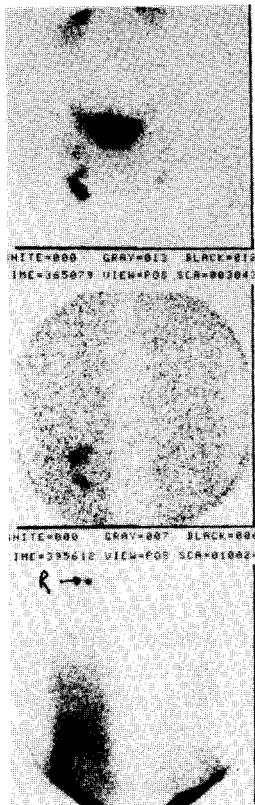


Fig. 3. Lymphatic scintigraphy of primary lymphedema of legs before lymphatic-venous shunt. In the right leg, tracer activity is seen in the lower portion, less clear in the mid-thigh with nodal visualization in the groin. On the left neither lymphatics nor nodes are detectable.

Fig. 4. Lymphatic scintigraphy of patient shown in Fig. 3 after lymphatic-venous shunt. In contradistinction to the preoperative studies both sides now show tracer activity not only in the lower legs but also in the thighs, and nodes are visualized in the groin and parailiac region.

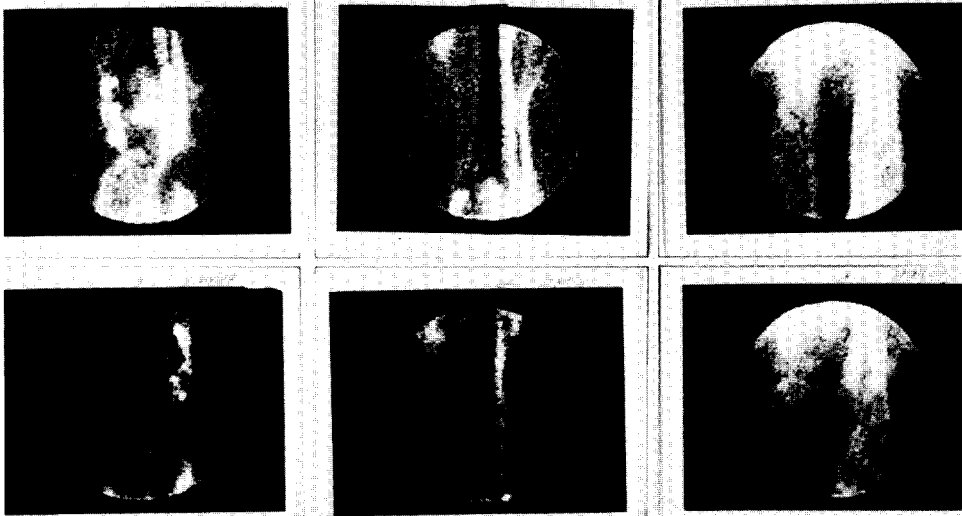


Table 3
Registry of Operations (Lymphatic Microsurgical)
(updated every 5 years)

LAL	Lymphatic ligation for "antigravitational" reflux
LNVA	Lymph nodal-venous anastomosis
LVA	Lymphatic venous anastomosis
LCVA	Lymphatic-capsular-venous anastomosis
LLA	Lymphatic-lymphatic anastomosis
SLAT	Segmental lymphatic autotransplantation
SLOT	Segmental lymphatic allotransplant
LVLA	Lymphatic-venous-lymphatic anastomosis

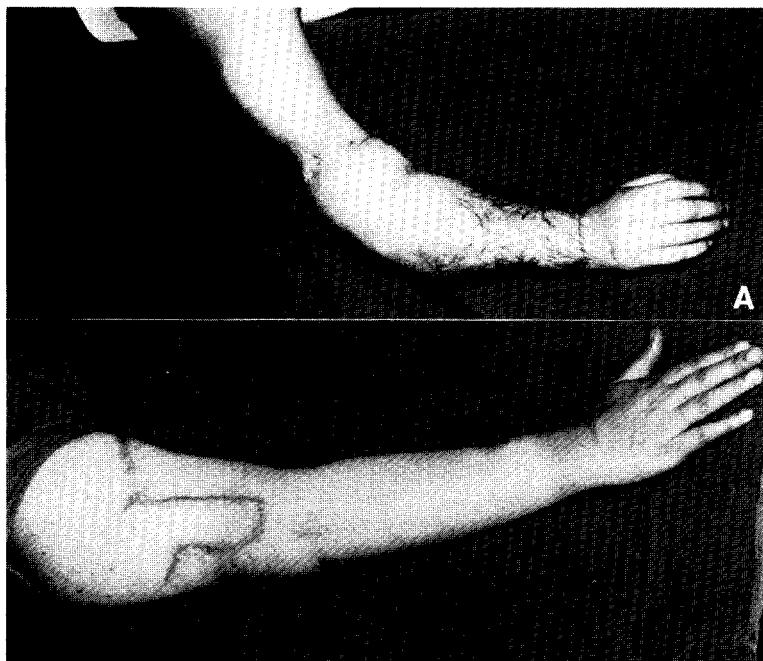


Fig. 5. Secondary lymphedema (obliterative lymphangitis) before (A) and after (B) plastic reconstruction.

where ipsilateral venous disease coexists with lymphatic disease, direct lymphatic shunts are contraindicated and alternative approaches such as lymphatic-lymphatic bypass (9) and interposition autologous vein grafts (i.e., lymphatic-venous-lymphatic angioplasty) (10) may be used. The latter technique has proved advantageous in that suitably long venous segments are readily harvested, several lymphatics can be anastomosed by invagination into relatively large caliber veins and the oper-

ative time is comparatively shorter (~2-3 hours).

In assessing outcome, a guideline for edema regression and function has been proposed (Table 2). More recently, lymphangioscintigraphy has been advocated (11) as shown before (Fig. 3) and after (Fig. 4) direct lymphatic-venous anastomosis. On the basis of extensive clinical experience over the last 18 years (Table 3), we are optimistic regarding the value of microsurgical shunts and plastic recon-

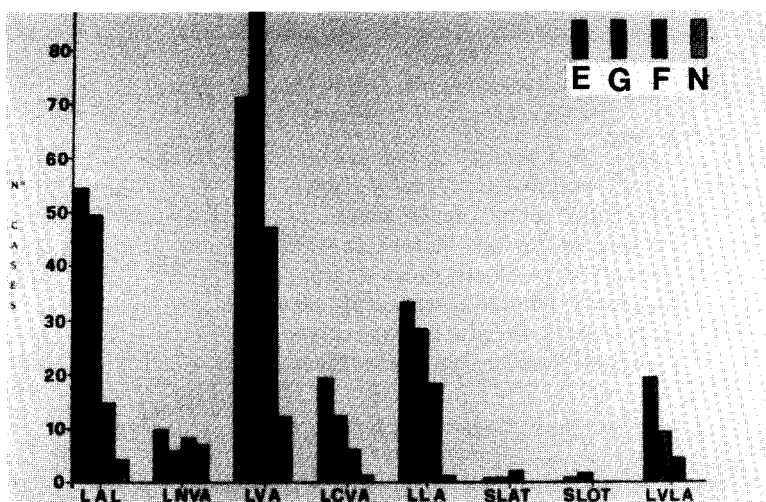


Fig. 6. Summary of lymphatic microsurgical operations and clinical outcome (see Table 2 for definition) /in our registry since 1973. LAL—lymphatic ligature for "antigravitational" reflux; LNVA—lymph nodal-venous anastomosis; LVA—lymphatic venous anastomosis; LVCA—lymphatic capsular venous anastomosis; LLA—lymphatic-lymphatic anastomosis; SLAT—segmental lymphatic autotransplantation; SLOT—segmental lymphatic allotransplantation; LVLA—lymphatic-venous-lymphatic anastomosis

struction in the management of peripheral lymphedema (Figs. 5,6). Inherent to this optimism is that proper diagnostic evaluation be done (5) and that each patient's condition, pathogenesis and etiology be taken into account before definitive treatment is proposed (12).

Although nonoperative therapy remains the cornerstone for treatment of lymphedema, it represents except in the early phase or in intermittent edema states only a partially satisfactory solution. Accordingly, with development of the operative microscope and microsurgical techniques for performing lymphatic shunt operations, greater enthusiasm has been generated to pursue these options. When based on careful diagnostic evaluation, consideration of the severity and persistence of the clinical condition as well as the etiology and pathogenesis of the lymphedema, these shunt techniques with little morbidity and no mortality hold promise of dramatically improving the largely intractable nature of these lymphedema disorders. With the refinement in imaging by isotope lymphography for validating long-term patency of these

operations, a whole new modality for safely examining the physiologic and clinical usefulness of these microsurgical lymphatic angioplastic operations is now available.

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