Lymphology 37 (2004) 182-184

GUIDELINES OF THE SOCIETÁ ITALIANA DI LINFANGIOLOGIA: EXCERPTED SECTIONS

C. Campisi, S. Michelini, F. Boccardo

University School of Medicine and Surgery (CC,FB), San Martino Hospital, Department of Surgery-Section of Lymphatic Surgery and Microsurgery, Genoa, Italy, and San Giovanni Battista Hospital (SM), Center of Neuro-Vascular Rehabilitation, Rome, Italy

Operative Treatment

Surgical techniques employed in the past to treat lymphedema would focus on bulk reduction of the affected limbs by a debulking-resection operation (cutolipofascectomy, total surface lymphangectomy, Thompson's operation, etc.). However, these were only symptomatic treatments: since they would not remove the cause of lymph flow obstruction, they were reducing lymphedema only temporarily, while they would require long hospitalization periods, and would frequently be accompanied by infections, delayed wound healing, loss of sensitivity, residual and progressing edema of the ankle and foot, as well as extensive retracting and disfiguring scars. Following the advent of Microsurgery, functional and causal therapeutic solutions for lymphedema were investigated and implemented aiming at draining the lymph flow or reconstructing the lymphatic pathways where they had been obstructed or were missing. Fine, repairing techniques were employed with direct intervention on the lymphatic structures. Microsurgery techniques have yielded positive and long-lasting results in the treatment of primary lymphedemas — including those in children — as well as in secondary lymphedemas following cancer treatment, involving lymph node resection in some 'critical' areas, such as in the armpit and the groin. Direct intervention on lymphatic-lymph node structures was first performed by multiple antigravitational ligatures of incompetent lymphatic and chyliferous vessels according to Servelle and Tosatti, to treat gravitational reflux lymphochyledema; Kinmonth (bridge) procedure was also employed, featuring the anastomosis of iliac-inguinal lymph nodes with an ileum segment, after mucosa removal from its mesenteric pedicle. However, with the progress made in surgical equipment design, armamentarium and techniques, two microsurgery methods have been developed for a "conservative and functional" treatment of lymphedema, namely derivative and reconstructive microsurgery. Derivative microsurgery techniques aim to resume lymph flow at the obstruction site, through a lymph-venous drainage in which lymph nodes or, directly, lymphatics are employed: Lymph node-Venous Anastomosis (LNVA), Lymphatic-Capsular-Venous Anastomosis (LCVA), End-to-end Lymphatic-Venous Anastomosis (EE-LVA), End-to-side Lymphatic-Venous Anastomosis (ES-LVA). Most recently, multiple, end-to-end and end-to-side lymphatic-venous anastomoses are most commonly employed, which are fashioned directly with the use of major veins or their collaterals, depending on the anatomic picture at the time of surgery, and performed at 1/3 midportion of the forearm volar surface

and in the inguinocrural region for the arm and leg, respectively. Conversely, with reconstructive microsurgery techniques, the lymphatic flow is resumed by overcoming the obstruction site either through a direct anastomosis of afferent and efferent lymphatics, or through the implant of autologous or venous segments between collectors down and upstream from the obstruction: Lymphatic-lymphatic Anastomosis (LLA), Segmental Lymphatic Vessel Autotransplantation (SLAT), Lymphatic-Venous- Lymphatic-Plasty or Lymphatic-Venous- Lymphatic Anastomosis (LVLA), Free Lymphatic-Lymph Nodal Flaps (FLF). With the LVLA technique, also bilateral lymphedemas can be treated, without risk of causing any iatrogenic lymphedema on the harvest site, as could instead happen when harvesting a lymphatic-lymph node specimen. Indications for the various microsurgical techniques depend on the presence of a viable lymphatic-venous pressure gradient in the affected limb. Should lymphostatic deficiency be associated with venous insufficiency (a condition mostly found in the lower extremities: varices, venous hypertension, valvular incontinence), derivative microsurgery is not recommended, while only reconstruction techniques can be applied.

Recommendation: Conventional surgical debulking-resective techniques are to be confined to cases in which it is necessary to remove excess skin and subcutaneous tissue of the lymphedematous limb, following a significant lymphedema reduction with CPT and/or microsurgery. Microsurgical procedures are highly beneficial especially in the early stages of disease: through the resumption of preferential lymph flow pathways in the affected extremity, good results (even healing) can be achieved with Microsurgery. Long term efficacy of lymphatic-venous anastomoses mainly depends on the accuracy of the adopted technique (the use of the operative microscope is essential) and on disease stage. Grade B.

GUIDELINES ON THE DIAGNOSIS AND THERAPY OF LYMPHEDEMA

ANGIODYSPLASIA AND LYMPHEDEMA

Cases of lymphatic dysplasia associated with vascular defects are defined as hemolymphatic malformations. According to the Hamburg classification (1988), congenital vascular malformations are grouped depending on the predominant defect: arterial, venous, lymphatic defects, A-V shunting defects or combined vascular defects. Each of these pictures is then subdivided into truncular and extratruncular forms, depending on the time and site of embryo defect onset.

Lymphatic malformations, classified under extratruncular (limited or diffuse) forms, are conventionally defined as lymphangiomas or lymphangiomatosis.

Truncular forms, which affect the major vessels (aplasia, hypoplasia, dilatation or hyperplasia), may cause lymphedema.

Further, lymphatic malformations may be associated with osteodystrophic syndromes (s.): angio-osteohypertrophic s. (with bone segment elongation), or angio-osteohypotrophic s. (with bone segment shortening). Comprehensive and integrated diagnostic procedures must be implemented with investigations of the arterial, venous, and lymphatic components. CT and MRI are useful to define malformation extent and relationships. Treatment features conservative medical-physical methods, in the mildest cases. Surgical treatment includes derivative and reconstructive lymphatic microsurgery, resection of tissues mostly affected by dysplasia, and antigravitational ligatures of incompetent lymphatics. Alternatively or in association with surgery, there are also treatment options by percutaneous sclerotherapy of lymphangiomatous and lymphangiectatic areas and/or embolotherapy of arteriovenous fistulas.

Recommendation: Hemolymphatic malformations, although rare, are due to highly complex vascular defects. They are nosographically classified according to the Hamburg Classification. Comprehensive and integrated diagnostic investigations must be conducted focused on the arterial, venous, and lymphatic components. CT and MRI are used to provide a comprehensive definition of malformation extent and relationships. Therapy features conservative, surgical, sclerotherapeutic methods, as well as percutaneous embolization, in varying mutual combinations, depending on the specific physiological features underlying each single case. Grade C.

NEONATAL LYMPHATIC DYSPLASIA

Lymphoscintigraphic investigations have recently been conducted on newborn babies with complex clinical pictures with hydrops, in order to determine the likely lymphatic origin of their malformation.

The task of CPR professionals in these cases is to conduct a primary assessment with treatment of respiratory and heart-blood circulation problems they are faced with from time to time, in order to ensure the survival of the baby, followed by a second, more accurate assessment, and by the final treatment.

Lymphatic circulation investigations by lymphoscintigraphy are part of the procedures of a secondary assessment. Indeed, from a physiopathologic point of view, if hydrops conditions are not due to congestive heart failure or to decreased osmotic plasma pressure and increased capillary filtration, they may well be due to lymphatic malformations (associated with chylothorax, chylous ascites, lymphedema, etc.)

Recommendation: In the assessment of a newborn baby with hydrops, after giving support to his/her life functions, also lymphatic circulation is to be considered as a possible cause of hydrops, an investigation which today is helped by lymphoscintigraphy.

Corradino Campisi, MD
Dpt. of General and Emergency Surgery
Lymphology and Microsurgery Center
Univ. of Genoa-Osp. San Martino
Largo Rosanna Benzi, 8
16132 Genoa, ITALY

(Full text available on ISL website)