LYMPHEDEMA REVIEW

Italian Guidelines on Lymphedema: New public regulations 2017

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Abstract Lymphedema in many countries is still considered an "aesthetic" problem, even though a chronic, degenerative, and debilitating disease. After a long period of study with internal and external experts, the Italian Ministry of Health has implemented National Care guidelines for lymphedema and other related disorders. This Document represents a fundamental element with regards to diagnostic and therapeutic regulation procedures that satisfy the health needs of these patients. In this paper the description of the main constituents of the document that has revolutionized the possibility of access the best specialized care facilities situated in the country.

Keywords Lymphedema, Public Health System.

General considerations

The problem of classifying treatment of primary and secondary lymphedema in both the public and private sectors, derives from a series of considerations. First of all the fact that the same global guidelines of the principal scientific societies that are experts in the subject (i.d. Consensus Document of the International Society of Lymphology) [1,2,3] illustrate the opinions of the leading world experts of the field: some consider lymphedema as an alteration of the lymph transport linked to intrinsic or extrinsic dysfunction of the lymphatic system itself; others consider it a disease. In fact the World Health Organization in the International Classification of Diseases (version number 9-ICD9) recognizes the various forms of lymphedema as diseases and not as symptoms or clinical manifestations. They were also assigned specific codification for the disease: 457.0 Breast cancer related lymphedema and 457.1 for all the other forms of primary and secondary lymphedema.

For these reasons in many countries patients with lymphedema find enormous difficulties coping with their health problems. The confusion and inadequate information, even among clinicians, with regards to the disease discredits even the scientific societies and associations of patients that struggle to be valid advocators for a serious confrontation with politicians who have to decide on funding treatment. As a result of this situation in almost all of the countries in the world a lymphedema patient is obliged to bear high health care costs that only a minority of patients can sustain.

Considering this particular critical situation, in Italy in 2006 a Commission was appointed by the Ministry of Health to draft ministerial guidelines on the management of national public health care for patients affected from lymphedema and other related diseases (Lipedema). The workshop was concluded in 2016 with the publication of a document that stipulates a series of specific health care regulations that guarantee free treatment under the

Universal Public National Health Care system for these patients.

The highlights of the Document, officially made public on Sep 15th, 2016, are the following:

Primary and Secondary lymphedema are considered a chronic disease, naturally progressive and degenerative, subject to clinical complications and exacerbations, similarly to other chronic diseases [4,5,6]

In Italy there will soon be a National Register that will document all the patients treated for both forms primary and secondary. It is currently estimated approx. 450.000 clinical cases nationwide with a majority of cases of the secondary form [7,8,9]

The primary form of lymphedema is considered in the new government document of the Essential levels of health and welfare assistance a "rare disease" therefore, these patients are identified with special classification codes that guarantee free specialized public health care treatment, from the clinical diagnostic staging to the treatment phase (therapeutic rehabilitation cycles, elastic garments), all of which are identified with specific codes that are issued by the National Health System.

Clinical complexity chart

Indicator name	N. of patients in day hospital setting	Setting	Rehabilitation medicine
Dimension	Process	Area	Volumes/Benefit
Data update	weekly/monthly/annually	Sources	Clinical records
Mathematical formula	N. of patients in treatment in DH/N. total patients seen with Lymphedema		
Expected standard	> 60%		
Regulation requirements			
Bibliography	*Although scientific literature does not provide evidence of a significant correlation between the activity volumes and the quality of the above procedures, it can be assumed that this relationship is a qualifying requirement for a Centre of Excellence*		

Indicator description
the indicator measures the N. of patients with a 'heavy' clinical picture being in charge to the Centre.
Indicator meaning
the indicator assesses the center's ability to manage complex patients.

Table I - Indicator of clinical complexity.

The primary forms of Lymphedema are included in the new Government Document for Essential Health Care Levels recognized in the category of 'rare disease' so that patients are identified with special classification codes that guarantees the patient specialized treatment, with respect to diagnostic testing clinical and instrumental, therapeutic cycles, as well as for the final elastic garments that are identified with specific codes by the National Health Care System. All forms of the disease are considered: present at birth (connatal), appearing within the first decade of life (early) or later onset (second, third, fourth, fifth, decade of life); these are the most common forms. Primary lymphedema can also be hereditary type (it is common that blood relatives are affected by the same disease), syndromic (this type of lymphedema is globally more complex, i.d. Klippel-Trenaunay Syndrome, the Ghoram Stout, the Noonan, the Hennekam ecc…) or sporadic (only one member of the family is affected and there is no presence of other blood relatives who have the same clinical problems) [10,11].

The diagnosis of lymphedema is clinical; in primary forms it is essential to perform a lymphoscintigraphic examination. Other tests (i.d. High resolution echography, videofluoroscopy, lymphangio nuclear magnetic resonance, echo color Doppler) contribute to a better diagnostic evaluation and therefore, a more accurate definition of treatment program. In the primary forms genetic testing is highly recommended to determine the presence and the type of mutation, also with regards to primary prevention for blood relatives of patient [12, 13].

Primary lymphedema (fig. 1) and Secondary (fig. 2), are subdivided, in accordance with the clinical stages indicated by the International Society of Lymphology, in 4 clinical stages:

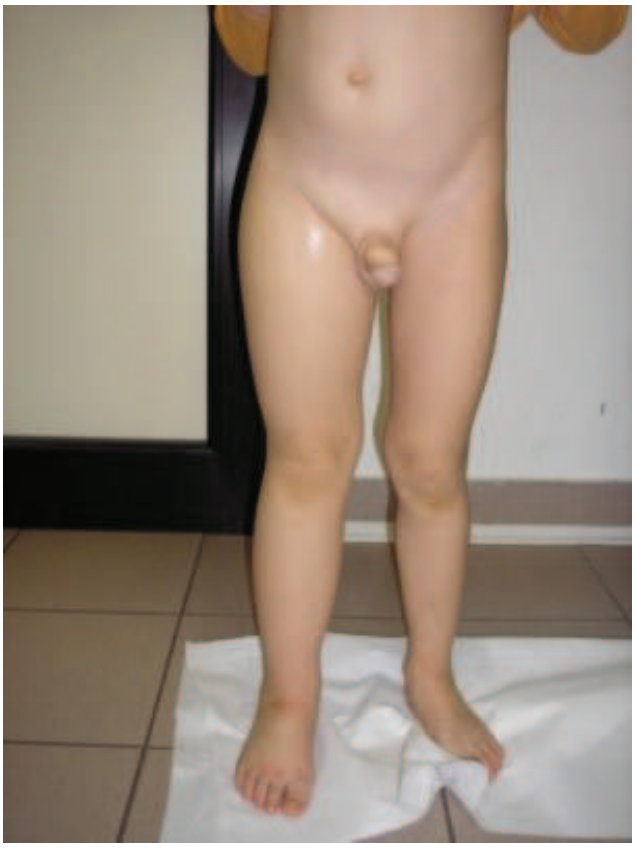

Figure 1 - Primary lymphedema of lower limb and genitalia.

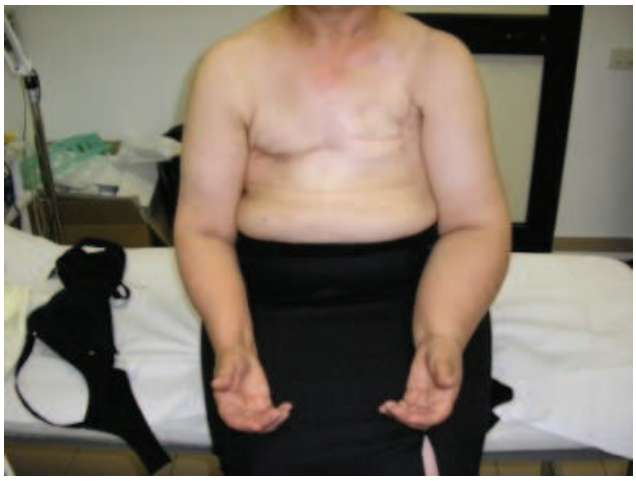

Figure 2 - Bilateral upper limbs secondary Lymphedema.

Stage 0: subclinical cases with possibility of developing edema (ex. Patients with mastectomy and axillary lymph node that present limbs apparently similar in volume and consistency).

Stage 1: presence of edema that shrinks partially with anti limb elevation and night rest;

Stage 2: uncomplicated elephantiasis

Stage 3: complicated Elephantiasis consisting of skin wounds, infectious, fungal, warty, ulcers, up until transformation linfosarcomatosa [3,4,7,10].

The National Health System provides treatment for these patients in various health care settings: outpatient services, day hospital or inpatient rehabilitation.

Outcome Indicator Chart			
Indicator name	N. of patients treated	Setting	Rehabilitative Medicine
Dimension	Outcome	Area	Volumes/Benefit
Data update	weekly/monthly/annually	Sources	Medical records
Mathematical formula	N. Lymphedema patients with clinical improvement (ROM, Barthel, Volume)/ total N. Lymphedema patients		
Lymphoscintigraphy	N. improved Patients/N. treated	Sources	Medical records
Expected standard		> 90%	
Normative requirements			

Indicator description
The indicator measures the N. patients improved with the received treatment
Indicator meaning
The indicator assesses the outcomes of the offered treatment and the fairness of procedures.

Table II - Outcome indicator.

The criteria that determines the care setting is essential based on the stage and the intensity of treatment after an accurate global evaluation of the needs of the patient taking into account also "social fragility", 'transportability' of patient and the necessity or not of medical assistance provided in an inpatient setting especially useful in the phase of the so called "attack Treatment"[14].

Every Italian Region has to organize regional observatory treatment Centers (Hub) which initially take into care and register patients in a Regional Register where periodically collected data is transmitted to the National Ministry of Health; the Center provides information and overall training programs.

The offered training programs are realized in collaboration with dedicated scientific societies. In these regional center it's possible to treat patients in 'outpatient', in Day Hospital, or 'inpatient' setting.

Each Region is requested to identify one or two reference hospitals for taking charge of clinical exacerbations or lymphangitis, ulcerative, trophic complications, even to the detriment of other organs or tissues. For maintenance treatment the organization provides various outpatient centers throughout the region to satisfy rehab needs of these patients. The treatment consists of combined physical therapy with at least manual lymphatic drainage, multilayer inelastic bandaging, physiotherapy under bandaging, ultrasound therapy and shock waves to be applied on fibrotic areas, linfotaping. Prescribed are 'monotherapy' (only lymph drainage, only physiotherapy etc.). Drug treatment is particularly indicated in early clinical stages of the disease: alpha and gamma benzopyrones constitute a therapeutic procedure that makes use of the proteolytic and pro-lymphokinetic and membrane stabilizers effects of such molecules. Surgical therapy uses techniques of reconstructive microsurgery and super-microsurgery; they must be carried out in highly specialized centers, by experienced staff certified by a recognized Masters degree, and are in synergy with the other treatment, since they cannot solve the problem in a definitive way in most cases. In case of lymphangitis, especially Erysipelas, is essential the use of broad-spectrum antibiotics (penicillin or macrolide), combined with anti-inflammatory drugs or cortisone. In cases (frequently in both primary and secondary forms) with repeated recurrence of infection it is recommended the use of penicillins for long periods (over the year), or desensitizing vaccines. The key issue is to control the obtained results by means the phase of major impact using physical treatment, by permanent elastic garment use (standard or 'tailored'); It is possible, to provide custom fit garments made of materials which permit the hold-up in certain anatomical regions in which the greater the tissue texture, major pressure effects by positioning 'thickness' that they are introduced in special 'pockets' pre-packaged [15,16,17,18,19].

They are provided, for patients with primary and secondary forms, elastic flat knitted garments that are replaced according to wear over time. These garments are identified with specific codes and are prescribable twice a year.

The Lymphedema reduces the quality of life; it is a disease that causes disability in patient. National Health System (in Italian Law 104/92) offers to the patients affected , and or family caregiver, special benefits (i.d. special work) permits to allow patients to take time off work to do treatment or medical visits and for caregiver to accompany patients, temporary paid leave from work to take care of a family member who is ill, reduction of taxes on certain essential items ecc [20].

A special role is reserved for specialized voluntary Associations that advocate to control the implementation at local level of the procedures provided and pay particular attention to the quality of provided services.

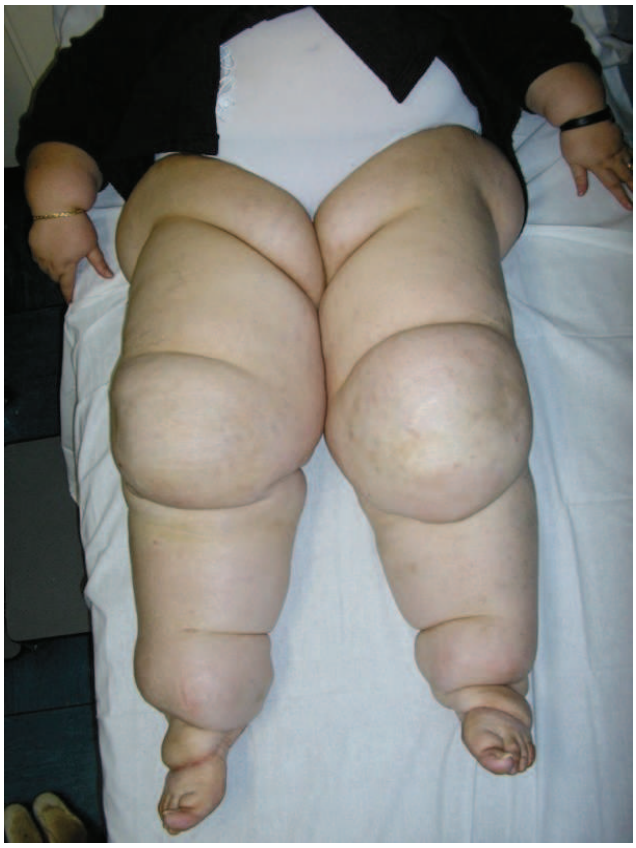

Figure 3 - Lower limb Lipedema.

The document also provides tools for the clinical and therapeutic outcome monitoring with specific scales that provide information on 'before' and 'after' treatment (Tables I and II).

Among the diseases related to Lymphedema, Lipedema (Fig. 3) is becoming more prevalent in the female population and often confused with Lymphedema, even by experienced clinicians. The disease affects the lower limbs; It can also be located in the upper limbs but spares always the distal portions of the limbs (hands and feet). The increase in volume is determined by the filing, under the skin, of large amounts of fatty tissue that does not respond to common adipose tissue self-regulation mechanisms and poorly to exercise. It is a stable clinical picture, not evolutionary, that does not come to infectious complications. It affects only women. It can give complications on the osteo-muscle-ligamentous structures (it is frequent Baker's cyst in the popliteal pit level for prolonged abnormal joint stress), and, lately, it can develop in Lipo-lymphedema for mechanical involvement of loco-regional lymph system. The treatment is absolutely symptomatic and aims to limit the mechanical obstacle and alleviate painful symptoms that frequently is one of the components of the clinical features [21, 22].

In view of an appropriate management of patients with lymphedematous disease, the following aspects are fundamental:

Recognized clinical experience of formal caregivers of the team (doctors, nurses and physiotherapists, orthopedic technicians, social workers);

Ability to work directly or with connected centers all the diagnostic tests needed to define the clinical picture (Lymphangioscintigraphy, high-resolution echography ultrasound, CT).

Possibility of execution, directly or by using approved genetic laboratory, the genetic test for the primary forms, exerting also through inter-regional agreements.

The very long (over ten years) and detailed discussion that has taken on the management of the disease, between Scientific Societies and patient Associations on the one hand and Government Structures on the other, has led to the final basic work for patients and as model for Health Care Systems, public and private, also of other countries. We hope that the final document produced and translated into legislation, inspired by international guidelines drawn up by leading experts, gives a basis for the resolution of support needs for about 300 million patients with primary and secondary lymphedema, identified by World Health Organization. With an adequate Training, incentive of professional caregivers and general practitioners, it will help pave the way to earlier diagnosis of the disease (with equally early Therapeutic management of the individual case) and improvement of primary and secondary prevention.

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