

IV Latin American consensus on the treatment of lymphedema

Authors: Jose Maria Pereira de Godoy, Mauro Andrade, Walter Ferreira Azevedo Jr., Raul Angel Beltramino, Eugenio Oscar Brizzio, Anacleto Carvalho, Jose Luis Cataldo, Jose Luis Ciucci, Henrique Jorge Guedes Neto, Claudia Stein Gomes, Solange Gomes, Maria de Fatima Guerreiro Godoy, Luis Daniel Marcovecchio, Delia E. Menaflor, Alejandro Latorre Parra, Lucia Wachawicz, Maria Bernadete Bottene, Anke Bergmann, Eliana Srur, Angel Esteban Guzmán, Tarso Túlio Nogueira, Roberto Simkin, Alfredo Jácomo, Ana Paula Rodrigues, Andréia Paula Kafajian- Haddad, Antonio Carlos D. Mayal, Delia Charles Edourard Otrante, Esther C. Aizic, Esther Azoubel, Maria Del Carmen J. Perez, Maria Estela Mancebo Vaz Martins, Walquiria C. Hueb, Aline Rodrigues, Melissa Morães, Monica Mayal, Carmem Nogueira, Daniela Mina Fukasawa, Eneida Couto de Mello, Cristina Hachul Moreno, Luis Gustavo S. Guedes, Anderson Gariglio.

Adress: Medicine School in São José do Rio Preto (FAMERP) av. Brigadeiro Faria Lima, 5416 Zipe code: 15090-000 - São Jose do Rio Preto-SP-Brazil

E-mail: godoyjmp@riopreto.com.br * author correspondence

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Abstract

In Latin America every three years for the last 12 years a group of professionals have been meeting to establish and arrive at a consensus on how to manage lymphedema. In 2011 the meeting took place in São Paulo, and brought together about 40 lymphologists, physiotherapists, occupational therapists and plastic surgeons. Several topics were identified as priorities by establishing the degree of consensus.

Key Words: Lymphedema, Consensus, Latin America

Introduction

Lymphedema is a problem that affects millions of people worldwide. Even so, it does not receive enough attention from governmental and private institutions in respect to prevention and treatment. Latin America is no different to other regions but every three years for the last 12 years a group of professionals led by Dr. Ciucci JL in Buenos Aires have been meeting to establish and arrive at a consensus on how to manage this disease. In 2011 the meeting took place in São Paulo, and brought together about 40 lymphologists, physiotherapists, occupational therapists and plastic surgeons. The literature stresses a need to improve concepts, diagnosis and management¹⁻³.

In this meeting several topics were identified as priorities by establishing the degree of consensus as follows:

Grade I when there was a consensus of all participants (strongly recommended)

Grade II when there is no consensus among all experts (medium recommendation)

Grade III when it is not recommended

The issues were divided in topics according to the established priorities:

1 - Concept, diagnosis, multidisciplinary treatment

2 - Anatomy, lymph drainage and new concepts

3 - Medications

4 - Lymphangitis and erysipelas

5 - Preventive aspects of lymphedema

6 - Interdisciplinary Treatment

- 7 - Pressotherapy
- 8 - Myolymphokinetic exercises and activities
- 9 - Manual lymph drainage
- 10 - Bandages in lymphedema
- 11 - Elastic compression in lymphedema
- 12 - Surgical treatment of lymphedema: lymphovenous anastomoses
- 13 - Foreskin and scrotal surgery
- 14 - Resection surgery

After the presentation of these topics, items were listed for each one to establish a consensus. Additionally, brief presentations of the main services in the different areas were presented on how they are managing each issue - however this was not part of the evaluation of consensus.

The concept of lymphedema

Lymphedema is an accumulation of water, salts, electrolytes, high molecular weight proteins and other elements in the interstitial space resulting from dynamic or mechanical changes of the lymphatic system which leads to a progressive increase of an extremity or body region with decreased functional and immune capacity, weight gain and morphological changes (Grade II).

Diagnosis of lymphedema

Regarding the diagnosis of lymphedema, the patient's history and physical examination were established as of fundamental importance (Grade I). However, it is suggested that specific minimum topics in the history must be investigated to standardize diagnostic criteria:

- Age at onset, infections, trauma, surgery, radiotherapy, trips to endemic areas, transient edema and family history
- Symptoms (pain as a synonym for complications or acute lymphedema)

Physical examination

Regarding the physical examination, investigation of the following topics is considered essential (Grade 1):

- Distribution of edema (unilateral, bilateral, often asymmetrical)
- Stemmer sign
- Godet sign (prognosis)
- Others (Hyperkeratosis, interdigital mycosis, clear or chylous vesicles, yellow nail)
- Measurement of perimeter and/or volume of limbs
- Body mass index (BMI)

Additional examinations

A Grade I consensus was reached in regards to complementary examinations, however specific indication of the main examinations was Grade II.

Lymphoscintigraphy as part of the initial assessment was given a Grade II recommendation in early diagnosis but Grade I in cases of diagnostic doubt.

Doppler ultrasound in the initial investigation was given Grade II recommendation, but when there was suspicion of lymphedema secondary to tumor, the recommendation was increased to Grade I.

Genetic testing in familial lymphedema was given grade I recommendation.

Anatomy of the lymphatic system

Anatomical aspects of the lymphatic system and the need to expand research were stressed in previous consensuses. The experience of the Caplan group and one of his disciples Dr. Ciucci JL has contributed to a better understanding of the anatomy of the lymphatic system. In this consensus, Dr. Ciucci presented new data and the suggestion that he should publish his recent findings in international scientific journals received Grade I recommendation. The most important recent findings are:

- An alternative route from the arm to the cervical lymph node chain was described
- An alternative route from the chest to the cervical lymph node chain was described
- That lymph in the deep lymphatic system of the legs can flow to the superficial system, which is different to the venous system
- There is an anatomical connection between the inguinal and axillary regions.

Medications

Drug therapy for prophylaxis and treatment of lymphedema was discussed and defined:

The term 'lymphoactive drug' was suggested (Grade I recommendation) for the category of substances that interfere in the lymphatic system.

The indication of lymphoactive drugs in the treatment of lymphedema received Grade II recommendation.

The prescription of lymphoactive drugs can be made at any time during (Grade I recommendation).

Among the drugs, micronized purified flavonoid fraction (MPFF - micronized diosmin and hesperidin) have a Grade I recommendation. However other lymphoactive drugs such as rutin, non-micronized diosmin, coumarin and calcium dobesilate and aminaphtone in cases of associated changes in permeability were given Grade II recommendation.

Some drugs with specific indications such as hormonal anti-inflammatory agents, antibiotics, antifungal

medication and analgesics can be used and received Grade I recommendation.

Prescription of diuretics was addressed in isolation due to the criticism of its use. Some experts reported using it in the early stages and in specific cases (Grade II recommendation).

Lymphangitis and erysipelas

Lymphangitis and erysipelas were addressed with specific topics being assessed such as:

The indication of prophylactic antibiotic therapy for lymphangio-cellulite associated with lymphedema received Grade I recommendation. Antibiotic therapy is recommended when patients with lymphedema are at risk of infections such as mycoses, trophic skin lesions and recurrent lymphangitis.

Among the medications reported, penicillin, first-generation cephalosporins and amoxicillin constitute first-line drugs. Other drugs such as erythromycin and clindamycin were mentioned. This therapeutic option received a Grade I recommendation.

The use of corticoids and non-hormonal anti-inflammatory medications were other drugs that were assessed and received Grade II recommendations.

Measures for the treatment and prevention of lymphedema were assessed and discussed, in particular, the necessity of having up-to-date knowledge and of spreading this knowledge to other key areas of the specialty. This aims to improve the safety of interventions, for example, in respect to the practice of lymph drainage (Grade I recommendation).

The improvement in surgical strategies to reduce aggression to the lymphatic system, such as the evaluation of sentinel lymph nodes is suggested, as well as other conducts in this respect. It is suggested (Grade I recommendation) that possible post-surgical fibrosis should be prevented and treated early and radiation should be avoided when possible.

A multidisciplinary and interdisciplinary approach in the treatment and prevention of lymphedema was discussed and its importance was recognized and given a Grade I recommendation. However, local, social and economic conditions of each treatment center should be considered and adapted to each situation.

The indication of pressure therapy was evaluated and received Grade II recommendation; it may be used at any time during treatment. It was suggested that pressure therapy reduces the volume (edema) of the limb (Grade I recommendation).

The recommended pressure is 40 mmHg (Grade II recommendation). The suggested time varies from 20 to 120 minutes (Grade II recommendation).

Myolymphokinetic exercises and activities

The main purposes of the use of myolymphokinetic exercises and activities were established (Grade I recommendation) and are:

- Reduction of edema
- Improve joint mobility
- Improve muscle trophism

The concept of myolymphokinetic activities and exercises was stressed where exercises correspond to programmed muscle activities and daily activities are not programmed (are performed randomly during the day including at work). The implementation of these activities, including in respect to the speed, intensity and repetition of movements, must be guided by a professional (Grade I recommendation).

Compression mechanisms exert a synergistic effect in reducing edema when combined with exercises and activities (Grade I recommendation).

The use of bandaging received Grade I recommendation in any phase of treatment, both decongestion and maintenance. Bandages can be substituted by compression stockings and sleeves when possible but this depends on the anatomy of the limb (Grade I recommendation).

The suggested technique for bandaging is multiple layers (Grade I recommendation).

A 30-40% and 50-60% elasticity was suggested for the bandages (Grade I recommendation).

It was suggested that the limb can remain under compression during the entire treatment period (Grade II recommendation).

Elastic compression in lymphedema received Grade I recommendation; this can be used in isolation (Grade I recommendation). However, the best result is when associated with exercise and muscle activity with a Grade I recommendation.

A compression of 30 to 40 mmHg was suggested for stockings and sleeves and given Grade I recommendation. One option was the use of two pairs of stockings when tolerated (Grade I recommendation).

Lymph drainage was recognized as a therapeutic procedure in the treatment of lymphedema (Grade I recommendation). Its use in isolation was given Grade II recommendation. However its use with other therapeutic modalities received Grade I recommendation.

The duration of treatment is variable and depends on the needs of each patient (Grade I recommendation).

The interval between sessions depends upon the patient's needs and can be flexible (Grade I recommendation).

Professionals indicated to perform lymph drainage must be qualified with specific knowledge of physiology, anatomy and pathophysiology and they must master an adequate technique (Grade I recommendation).

Contraindications

Absolute:

Absolute contraindications include active neoplasms in the region of drainage, active infections in the limb, heart failure, hepatic or renal decompensation and acute phase deep vein thrombosis (Grade I recommendation).

Surgery

Lymphovenous anastomosis is indicated when clinical treatment fails, when there is deterioration identifiable by lymphoscintigraphy and when the anatomical conditions are favorable (Grade II recommendation). The suggested technique is a derivative lymphatic-venous microsurgery (Grade I recommendation).

The specific preoperative complementary examinations are lymphoscintigraphy and duplex Doppler ultrasound (Grade I recommendation).

Their use for postoperative control was given a Grade II recommendation.

The maintenance of physical therapy or elastic compression is suggested and received a Grade I recommendation.

Indication of prophylaxis treatment for lymphedema in breast cancer received Grade II recommendation.

Foreskin and scrotal surgery

Foreskin and scrotal surgery in advanced cases of lymphedema was recommended along with physical treatment with manual lymph drainage and compression (Grade I recommendation).

Resection surgery

The main surgeries performed are partial dermolipectomy, Servelle's operation, Charles operation (Grade I recommendation).

Indicate for giant lymphedema with functional alterations involving the quality of life and after clinical treatment to remove remaining tissues (Grade I recommendation).

Amputation should be used in exceptional cases (Grade I recommendation).

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