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E.S.L.
European Society of Lymphology

46th ESL Congress European Society of Lymphology

June 16th-18th, 2022 - Assisi, Italy

Congress Venue

Domus Pacis
Santa Maria degli Angeli

Congress President
Marina Cestari

Scientific Coordinator
Sandro Michelini

European Society of lymphology

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Presidental Welcome

Dear Friend Lymphologist,

we have just lived an inedited sanitary emergency, COVID-19 pandemic. This situation has conditioned many aspects of our life, health, family affections, work and the world economy. In the same way, the conference activities have also been interrupted for long.

Although we are at the beginning of a possible return to an almost normal situation, the President of the Congress, together with the Scientific Coordinator, myself and the members of the Executive Committee have courageously decided to set the new dates of the congress on June 2022. We all intensely desire to be able to meet again, to exchange our scientific opinions, to stay together even on convivial occasions and to continue the activity of our society, with the aim above all to update ourselves on new scientific researches in the lymphological field, stimulating the participation of young lymphologists, and expand lymphological knowledge all over Europe.

The Congress of the European Society of Lymphology 2022, which Marina and Sandro are planning with considerable organizational efforts will certainly be an opportunity for the growth of the ESL under different points of view. In the meanwhile, the activity of the Society and the interest in the European Lymphology have been definitely live and dynamic.

Finally, I am really pleased to welcome you all to our meeting and hoping in a return to normal as quickly as possible.

Francesco Boccardo

ESL President

Dear Colleagues,

after two years of postponements, I am pleased to finally welcome you to the 46th Congress of European Society of Lymphology taking place in Santa Maria degli Angeli - Assisi on the 16-18 of June 2022.

The aim of the Congress is to contribute to the progress in the field of Lymphology through the scientific programme and will include lectures and free paper sessions as well as workshops and poster session. I thank you for your updated contributions on new research in the lymphological field.

I hope you will enjoy the Congress and the city of Assisi, a UNESCO world heritage site full of history, art and spirituality

Marina Cestari

Congress President

General Information

The ESL CONGRESS 2022 will be held as an Event "BLENDED":

- *in-person* at SANTA MARIA DEGLI ANGELI-ASSISI on JUNE 16-18, 2022
- *virtual* for participants unable to participate in person

CONGRESS VENUE

DOMUS PACIS - Piazza Porziuncola, 1 - 06081 Santa Maria degli Angeli - Assisi (PG) Italy

Phone: 0039 075/80 43 530 - Website: www.domuspacis.it

ABSTRACTS

Abstracts will be published on the "European Journal of Lymphology" and distributed at the Congress

WEB SITE: www.eurolymphology.org

HOTEL ACCOMMODATION

The Organizing Secretariat has reserved some available rooms in FULL BOARD in the Congress Venue at the agreed prices:

- Double room single use **euro 85,00** per person per day
- Double room **euro 75,00** per person per day

Please add also **euro 1,50** per person per day as city tax (it is mandatory)

For further information and reservation please contact Mrs. Daniela Carletti: segreteria2@gccongressi.it

TRAIN CONNECTION ROME-ASSISI-ROME

There are trains connecting Rome Airport-main Railway Station-Santa Maria degli Angeli-Assisi (PG).

The train schedules can be found on the link: www.thetrainline.com where you can also buy your tickets or ask the Organizing Secretariat for any further information: segreteria2@gccongressi.it

UMBRIA INTERNATIONAL AIRPORT SAN FRANCESCO D'ASSISI

Please follow this links: www.adr.it (Rome FCO APT) or www.airport.umbria.it (Perugia APT).

Here you will find all information on destinations, flight timetables, ticket office, online check-in, offers, parking, car rental and travel services.

SIMULTANEOUS TRANSLATION

An English versus Italian simultaneous translation service will be provided

REGISTRATION FEES

	IN PERSON	VIRTUAL
ESL Members *(up-to-date with 2022 ESL membership fee)*	**FREE**	**FREE**
No Members	**€ 200,00** *(22%VAT included)*	**€ 122,00** *(22%VAT included)*
Social Dinner	**€ 50,00**	

To benefit from free registration fee, ESL Members must have settled their membership fee 2022

REGISTRATION FEE INCLUDES

- Admission to the congress area, scientific sessions
- Congress kit and certificate of attendance
- Abstracts of the Congress
- Simultaneous translation English versus Italian
- Coffee- breaks

Lunches on Friday 17th and Saturday 18th June are available only for those staying at the following hotels: Hotel Domus Pacis, Hotel Antonelli, Domus Benedicta, Hotel Donnini, Hotel Frate Sole

Payment of ESL annual membership fee

BNP Paribas Fortis
Beneficiary Bank Account: **European Society of Lymphology**
IBAN: **BE44 00185112 3445**
BIC: **GEBABEBB**

Please send copy of the payment to
G.C. CONGRESSI: andreina.mancini@gccongressi.it

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Please send copy of the payment together with the registration form to
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SOCIAL DINNER:

The Social Dinner will be held on Friday 17th
Those wishing to participate must make a reservation at G.C. Congressi having paid the corresponding fee to:

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ESL WEB SITE:

www.eurolymphology.org
please visit it for advanced versions of the programme

HOW TO BECOME AN ESL MEMBER:

FULL MEMBER with the rightto vote

- Present a report to the ESL Congress 2022
- Send a CURRICULUM VITAE to the President of the ESL Society: prof. Francesco M. Boccardo ***francesco.boccardo@unige.it***
- Your CV will be presented to the ESL Executive Committee during the ESL Congress 2022. Once approved you will be asked to pay your 2022 membership fee of **euro 80,00***

ASSOCIATE MEMBER with no right to vote

- Just Send a CURRICULUM VITAE to: ***francesco.boccardo@unige.it***

**Payment of ESL annual membership fee*

BNP Paribas Fortis
Beneficiary Bank Account: **European Society of Lymphology**
IBAN: **BE44 00185112 3445**
BIC: **GEBABEBB**

ORGANIZING SECRETARIAT

GESTIONE CONGRESSI srl
Via Pietro Borsieri, 12 – 00195 Rome – Italy
e-mail: segreteria@gccongressi
andreina.mancini@gccongressi.it

HOTEL INFO

Attention: **Mrs. Daniela Carletti**
e-mail: segreteria2@gccongressi.it

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Thursday, June 16th, 2022

15:00-15:30 **REGISTRATION**
15:30-16:00 **OPENING CERIMONY**
Opening Lecture
Pissas A. (Montpellier-FR)
16:00-17:30 **SESSION I: IMAGING**
Chairs: Baumeister R., Bourgeois P., Macciò A.
Near-Infrared Fluorescence Imaging in Chylous Diseases
Amore M., Castro P., Salvia S, Gerez N., Bengoa G., Marcovecchio L. Buenos Aires-AR
"Natural History" of dermal backflows? Insights from lymphoscintigraphic investigations and practical recommendations
Bourgeois P., Callebaut G. Brussel-BE
The investigations of the deep lymphatic system of the lower limbs: spect-ct is mandatory!
Bourgeois P., Callebaut G. Brussel-BE
The popliteal lymph nodes and the investigations of the superficial lymphatic system of the lower limbs: spect-ct is recommended!
Bourgeois P., Callebaut G. Brussel-BE
Lymphoscintigraphic pattern in CEAP 0s: role of deep lymphatic circulation
Macciò A. Savona-IT
Can lymphoscintigraphy be part of the third millennium nuclear medicine?
Mango L. Rome-IT
Study of the superficial and deep lymphatic compartments of the limbs: the contribution of lymphoscintigraphy
Villa G., Pennone M., Boccardo F., Donegani I., Miceli A., Campisi C. C. Genoa-IT
17:10-17:30 **DISCUSSION**
17:30-18:00 **LECTURE**
Compression possibilities in lymphedema
Chair: Boccardo F.
Schingale F. J. Pommelsbrunn-DE
18:00-19:30 **SESSION II: PEDIATRIC LYMPHOLOGY**
Chairs: Cestari M., Leone A., Moffatt C.
Pediatric Lymphedema: General View From Diagnosis to Management
Borman P. Ankara-TR
The true story of the real lymph I
Braun D., Papendieck C. M. Buenos Aires-AR
The true story of the real lymph II
Papendieck C. M. Braun D. Buenos Aires-AR
Lymphoedema in development age: primary lymphoedema treatment
Di Niccolo R. Bari-IT
Diagnosis and therapy of lymphedema in childhood; family involvement in therapies.
Leone A. Catania-IT
FLT4/VEGFR3 mutations are associated with segmental lymphatic dysfunction and initial lymphatic aplasia in patients with Milroy disease
Ningfei Liu, Minzhe Gao Shanghai-CHN
Dysfunction of dermal initial lymphatics of the arm and upper body quadrant causes congenital arm lymphedema
Ningfei Liu, Minzhe Gao, Ziyou Yu Shanghai-CHN
Complex decongestive therapy in children with lymphedema
Rovnaya A. St Petersburg-RU
19:20-19:30 **DISCUSSION**
16:30-17:30 **WORKSHP ON VASCULAR ULCERS**
Macciò A. Savona- IT

Friday, June 17th, 2022

07:30-08:30 **ESL EXECUTIVE COMMITTEE**
08:30-11:00 **SESSION 3: PHYSICAL THERAPY**
Chairs: Hamadé A., Moneta G., Szuba A.
Impact of elastic decompression taping methodology modulating phasic mechanical forces on collecting lymphatic contractions
Blow D. Rome-IT
Kinesiotaping in Lymphoedema
Borman P. Ankara-TR
Analysis of data extracted from Linforoll in Patients with secondary lymphoedema in the period 2014-2018
Caldirola R., Cascino E., Olszewsky W. L., Zaleska M., Cestari M. Lecco-IT, Rome-IT, Terni-IT; Warsaw-PL
The comprehensive treatment of giant lymphoedema
Manokaran G.– Chennai-IN
Effectiveness of Complete Decongestive Therapy Phase I (CDT) for the treatment of Lymphoedema
Dimakakos E., Michopoulos E., Papathanasiou G., Krousaniotaki K., S. Michopoulo S., Profka A Athens-EL
The physiotherapist's competences with respect to the " Italian Ministerial Guidelines on Lymphedema and other Pathologies related to the Lymphatic System" and the " 2016 Consensus Document" of the International Society of Lymphology
Galli T. Milan-IT
The place of graduate degressive compression socks in amateur athletics affected by lymphoedema of lower limbs
Hamadé A., Blanquier G., Finiels Q., Baumann A, Michel P., Lehn-Hogg M., Buschenrieder N., Woehl B. Mulhouse-FR
The Effects of Trunk Stabilization Exercises on Body Endurance and Pain in Lipedema Women
Kostanoglu A., Demircan G. İstanbul-TR
Oedema of lower limbs in great obese patient: problems to solve waiting bariatric surgery
Moneta G., Nesci A., Michelini S., Failla A. Rome-IT
Is it logical to treat intensively patients with lymphoedema in an outpatient structure
Pissas A., Gras O., Scaratto M., Gallon F. Montpellier-FR
Self-Management with Compression in Lymphoedema
Schingale F. J. Pommelsbrunn-DE
Benefit of an adjustable compression system (coolflex/coolflex boot) for the reduction of edema (akrö study)- preliminary results
Szuba A. Faerber G., Kahle B., Hirsch T. Wroclaw-PL, Hamburg-DE, Lubeck-DE, Halle-DE
High force 3 x 30 min compression method of foot and calf removes most edema fluid and enables immediate usage high compression garment
Zaleska M., Olszewski W. L. Warsaw-PL
10:40-11:00 **DISCUSSION**
11:00-11:30 **COFFEE BREAK**
11:30-12:30 **WORKSHOP ON: PARTICULARITIES OF GENITAL LYMPHEDEMA: FROM DIAGNOSIS TO TREATMENT**
Chair: Michelini S.
11:30-11:50 **What do we know about the patient profile and the diagnostics specifics**
Borman P.Ankara-TR
11:50-12:10 **New perspectives in genital lymphedema management**
Mestre-Godin S. Montpellier-FR
12:10-12:30 **DISCUSSION**
12:30-13:30 **SESSION 4: COMPLICATIONS AND PREVENTION**
Chairs: Amore M, Johansson K., Macciò A.
Sirolimus Induced Lymphedema
Amore M. Buenos Aires-AR
Early intervention with compression Garments prevents progression in mild breast cancer-related arm lymphedema: a randomized controlled trial
Johansson K., Blom K., Nilsson-Wikmar L., Brogårdh C. Lund, Stockholm-SE
The benefit of no, immediate or delayed compression garments to prevent progression in mild breast cancer-related arm lymphedema: a one-year follow-up.
Johansson K., Blom K., Nilsson-Wikmar L., Brogårdh C. Lund, Stockholm-SE
Lymphatic ulcers: how photobiomodulation help us?
Macciò A. Savona-IT
High level of lipids retained in stagnant tissue fluid/lymph may be responsible for adipocyte hypertrophy and hyperplasia in lymphedema
Zaleska M., Olszewski W. L., Miller N. Warsaw-PL, Cambridge-UK
13:20-13:30 **DISCUSSION**
13:30-14:30 **LUNCH**
14:30-15:00 **Lipoedema: the new horizon from International Consensus Document by International Lipedema Association**
Corda D., Pavia-IT - Hardy D., Kendal-UK
15:00-17:00 **SESSION 5: LIPEDEMA**
Chairs: Cornely M., Michelini S., Onorato A.
Usa Lipedema Guidelines
Herbst K., Tucson-AZ-USA
Lipedema: usefulness of 3D Ultrasound diagnostics
Cestari M., Terni-IT
Joint hypermobility is more frequent in Lipedema than in Lymphoedema patients
Forner-Cordero I., Vázquez-Díez J., Muñoz-Langa J. Valencia-ES
Measurement and assessment of Lipedema
Onorato A., Filelfi S., Ure C., Cornely M. Udine-IT, Cologne-DE
What are the most important features of Lipedema?: Clinical results from a prospective cohort study.
Forner-Cordero I., Muñoz-Langa J. Valencia-ES
Lipohyperplasia dolorosa or Lipoedema. Which term is better for this condition?
Cornely M. Cologne-DE
Comparison of the effectiveness of low-carbohydrate, high-fat diet with moderate-carbohydrate and fat diet in reducing body weight and body composition parameters in patients with Lipedema
Szuba A., Jeziorek M., Regulska-llow B. Wroclaw, Poland
Onset of Dercum's disease (adiposa dolorosa) following COVID-19 infection
Iker E., Santa Monica–CA-USA
ESL Consensus Document on Lipedema: Proposal for discussion
Michelini S., Boccardo F., Cestari M. Marino, Rome-IT, Genoa-IT, Terni-IT
16:40-17:00 **GENERAL DISCUSSION**
17:00-18:00 **OPEN COFFEE BREAK**
17:00-19:00 **SESSION 6: DIAGNOSIS**
Chairs: Ricci M., Zalewska M. T., Wald M.
Molecular pathways involved in primary lymphedema
Bertelli M., Bonetti G., Paolacci S., Samaja M., Maltese P. E., Michelini S., Michelini S. jr, Michelini Silvia, Ricci M., Cestari M., Dautaj A., Medori C. Rovereto, Brescia, Rome, Ancona, Terni IT
National Register of Rare Diseases, cases with rare diseases attention code RGG020 from 2017/2018, experience of the Ligurian Center and comparison with national data.
Eretta C., Bianchi C., Fabbricotti A., Beatini L., Colombini M., Cadeo A., D'Ambra L., Rodriguez Antinucci V., Pigoni A., Isaia S., Arfanotti D., Berti S. Sarzana-IT
Circumferential measurements to calculate lower limb volume in persons with lymphedema. What segment length is to be recommended
Johansson K., Jönsson C, Bjurberg M., Brogårdh C., Lund-SE
Perometric 3D scan in the management of chronic Lymphedema
Macciò A. Savona-IT
Immunohistochemical observation of the microvasculature of the human mammary tissue
Okada E., Toyama- JP
Regional lymphedema - the lymphatic drainage system may be damaged locally in various lower limb regions but not necessarily in the whole limb
Zaleska M., Olszewski W. L. Warsaw-PL
ICF and Disability
Ricci M. Ancona-IT
Lymphology - Origins and Future Development?
Wald M. Prague-CZ
Novelties in genetic stratification of lymphedema
Vikkula M. Bruxelles-BE
18:30 **DISCUSSION**

15:00-17:00 **SESSION 7: POSTER**

Chairs: Boccardo F., Cestari M.

1. THE EFFICACY OF REHABILITATIVE AND WOUND HEALING APPROACHES IN A MORBID OBESE PATIENT WITH LYMPHEDEMA
Civelek G. M., Borman P., Köse Yılmaz N., Sahbaz Pirincci C., Ulusoy S., Özdemir E. E. Ankara-TR

2. CONGENITAL LYMPHEDEMA IN A PATIENT WITH PROTEIN C DEFICIENCY AND HYPOXIC BRAIN INJURY
Civelek G. M., Dalyan M., Sahbaz Pirincci C., Celik C. Ankara-TR

3. LYMPHATIC CORD FOLLOWING AXILLARY NODE DISSECTION
Johansson K., Chong H., Ciornei C. D., Brorson H., Mortimer P. Lund, Malmö-SE, London-UK

4. CELLULITIS (DERMATO-LYMPHANGIO-ADEMITIS, DLA) IN LIMB LYMPHEDEMA – ANTIBIOTIC ACUTE AND CHRONIC THERAPY, PROPHYLAXIS
Olszewski W. L., Zaleska M., Swoboda E., Stelmach E. Warsaw-PL

5. THE LOWER LEG CONFINED INFLAMMATION IS GRADUALLY FOLLOWED BY DAMAGE TO LYMPHATICS AND DEVELOPMENT OF WHOLE LIMB LYMPHEDEMA
Olszewski W. L., Zaleska M. Warsaw-PL

6. LYMPHATIC COMPONENT OF LEG VENOUS ULCERS – LYMPHOGRAPHY EVALUATION OF THE SIZE OF PERIVENOUS INFLAMMATION
Olszewski W. L., Zaleska M. Warsaw-PL

7. LONG-TERM FOLLOW-UP OF SILICONE TUBE IMPLANTATION FOR BYPASSING THE SITE OF LYMPH FLOW OBSTRUCTION - 2020
Olszewski W. L., Zaleska M., Hydrabadi R., Banker M., Kurkular P. Warsaw-PL, Ahmedabad-IN

8. LYMPHEDEMA MAY BE CAUSED NOT ONLY BY LYMPHATIC OBSTRUCTION BUT ALSO BY LOSS OF LYMPHATIC CONTRACTILITY
Olszewski W. L., Zaleska M. Warsaw-PL

9. PECTORALIS MAYOR RUPTURE IN A PATIENT WITH BREAST CANCER. SECONDARY LYMPHEDEMA. A CASE REPORT
Gil Hernández M.S., Crespo Cobo M.P., Martínez Serrano S. Madrid-ES

10. MANAGEMENT OF GENITAL LYMPHOEDEMA
Manokaran G. Chennai-IN

11. BREAST CANCER-RELATED LYMPHEDEMA: RISK FACTORS AND PREDICTION MODEL
Martínez-Jaimez P., Armora Verdú M., Forero C., Álvarez Salazar S., Fuster Linares P., Monforte-Royo C., Masia J. Barcelona-ES

12. ADJUSTABLE COMPRESSION WRAPS SYSTEMS IN SEVERE LIPOLYMPHOEDEMA
Crespo Cobo M. P., Gil Hernandez M. S., Hurtado Lopez M. D. Madrid-ES

13. MORPHOLOGICAL CHARACTERIZATION OF ADIPOSE TISSUE FROM PATIENTS AFFECTED BY LIPEDEMA
Michelini S., Greco S., Vaia N., Puleo V., Pellegrino P., Giordano A., Michelini S.jr, Herbst K., Ciarmela P., Cinti S Rome, Ancona-IT, CA-USA

14. DESIGN OF A MULTIDISCIPLINARY PREVENTIVE INTERVENTION FOR BREAST CANCER-RELATED LYMPHEDEMA: AN INTERNATIONAL CONSENSUS
Martínez-Jaimez P., Fuster Linares P., Masia J., Monforte-Royo C. Barcelona-ES

15. IMPACT OF LOCKDOWN ON PPL IN ITALY: FOCUS ON THE EFFICACY OF EDUCATIONAL LYMPHOEDEMA CHILDREN'S CAMPS IN DAILY SELF MANAGEMENT
Stasi E., Cioffi M., Baldovino S., Sciascia S., Moffatt C., Quere I., Roccatello D. Turin-IT, Nottingham-UK, Montpellier-FR

20:30 **SOCIAL DINNER**

Saturday, June 18th, 2022

08:30-10:00 **SESSION 8: CLINICS**

Chairs: Borman P., Campisi C. C., Pissas A.

Fat deposition as a result of inflammatory response in experimental lymphedema
Andrade M., Cuadrado G., Ariga S., Lima T., Souza H. São Paulo-BR

The comparative frequency of breast cancer-related lymphedema determined by perometer and circumferential measurements: relationship with functional status and quality of life (Preliminary Report)
Civelek G. M., Borman P, Sahbaz Pirincci C., Uncu D., Uçar G., Yaman A., Dalyan M. Ankara-TR

Evaluation of Sleep Quality, Depression and Quality of Life in Patients with breast cancer related Lymphedema
Civelek G. M., Akinci M. G., Dalyan M. Ankara-TR

Lymphatic Vessel genital oedema: Scrotal lymphoedema and preputial elaphantiasis: case report
Sciuscio M., Lecce-IT

Lymphatic vessels senescence: current evidence and implications for clinicians
Onorato A., Filelfi S., Ure C., Goswami N. Udine-IT, Wolfsberg, Graz-AT

A questionnaire based survey to assess the awareness amongst health care professionals about breast cancer-related lymphedema.
Vagal M. Mumbai-IN

9:30-10:00 **DISCUSSION**

10:00-10:30 **COFFEE BREAK**

10:30-13:00 **SESSION 9: SURGERY**

Chairs: Boccardo F., Brorson H., Campisi C., Masia J.

Microsurgical treatment of breast cancer-related Lymphedema combined with complex physical therapy: preliminary results of the effect on arm volume change
Adrianssens N., Harnie S., Vandyck E., Van Rillaer S., Swaelen A., Hamdi M., Zeltzer A. Brussels-BE

Long term outcome of LYMPHA technique after 10 years' follow-up
Boccardo F., Santori G., Villa G., Accogli S., Dessalvi S. Genoa-IT

Pros and cons of the different microsurgical techniques proposed today for the treatment of lymphedema
Boccardo F. Genoa-IT

Liposuction Normalizes Lymphedema Induced adipose tissue hypertrophy in elephantiasis of the leg-How long do outcomes last?
Brorson H., Ohlin K., Svensson B., Hoffner M., Karlsson T. Lund- Malmö-SE

From lymph to fat - Lessons learned during 25 years of complete reduction of arm lymphedema
Brorson H., Ohlin K., Svensson B., Hoffner M., Karlsson T. Lund- Malmö-SE

Staging-guided and long-lasting surgical treatment of peripheral lymphedema – The GALT System (Genoa Advanced Lymphedema Therapy)".
Campisi C. C., Campisi C. Genoa-IT

Microlymphatic pumps for longstanding lymphedema
Baumasteir R., Weiss M., Frick A., Wallmichrath J. Monaco-DE

Long-term effects of surgical treatment of secondary lymphoedema. Do I still need complex decongestive therapy afterwards?
Cornely M. Cologne-DE

Long Term Patency of MLVA
Dessalvi S., Santori G., Villa G., Accogli S., Boccardo F. Genoa-IT

Surgical treatment of post-mastectomy in advanced lymphedema through the super charge submental lymphatic transfer
Fornasier F., Aparicio Alcazar J. J., Umaña M. F. Milan-IT, Alicante-ES

Microsurgical treatment of lymphedema
Gentileschi S. Roma-IT

12:20-12:40 **DISCUSSION**

12:40-13:00 **FINAL REMARKS**

13:00-14:00 **ESL GENERAL ASSEMBLY**

14:00-15:00 **LUNCH**

14:00-15:00 **ASSISI TOUR**

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Acknowledgement

Thursday, June 16th, 2022

Session 1: Imaging

Chairs: Baumeister R., Bourgeois P., Macciò A.

OPENING LECTURE

PISSAS A., *Montpellier-FR*

GEL /ESL (1979-2022) : A MARVELOUS ADVENTURE

On behalf of Pr Albert LEDUC honorary president I have the mission to share with all of you the happiness of the creation, the history, the transformation of our society: from its initiation in 1979, its creation in 1980. I shall try to tell you the difficulties, the interrogations, the success of GEL, the change of name ESL in 1996. Many challenges: the rules and by-laws, the languages, the position with ISL. Of course it existed clear competition between some colleagues; but from detrimental this competition became a boon... This transformation and the link with ISL were possible through women and men under the auspices of brotherhood, the mutual respect, the adaptation of rules, and through 45 congresses which represented a real moment of happiness .

The history is never finished and new generations will write the following. Some guidelines could be proposed. I shall propose to our president F. BOCCARDO and to S. MICHELINI in charge of website to introduce this presentation.

NEAR-INFRARED FLUORESCENCE IMAGING IN CHYLOUS DISEASES

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Background: Near-Infrared (NIR) Fluorescence imaging is increasingly gaining intraoperative applications and it can provide more anatomic and functional information of the chylous - lymph system.

Aim: Is to evaluate the useful of NIR imaging to diagnosis and treatment of chylous diseases.

Material & Methods: We evaluated 5 (N=5) patients with chylous disroders. 3 (N=3) primary and 2 (N=2) secondary, after surgery. We perform ICG lymphography by trans nodal inguinal injection and Intraoperative – Nasgogastric Tube with ICG + cream injection in all cases, ICG stomach injection by preoperative endoscopy in N=1, ICG + cream oral infusion 12 hours preoperative in N=1. In all cases we performed fluorescent guided laparoscopic surgery.

Results: With the guidance of real-time fluorescence lymphography, the leaks were identified and treated in all patients and chylous reflux was managed successfully by surgical interventions.

Conclusion: Near-infrared fluorescence imaging with ICG provided highly sensitive and real-time imaging of lymph-chylous disorders.

"NATURAL HISTORY" OF DERMAL BACKFLOWS? INSIGHTS FROM LYMPHOSCINTIGRAPHIC INVESTIGATIONS AND PRACTICAL RECOMMENDATIONS

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Background: Vascular lymphatic reflux (VLR), also called Dermal Backflows (DBF), are frequently seen in patients with Lower Limb Lymphatic Edema (LLLE) in the framework of the lymphoscintigraphic investigations (LLLySc) of these situations. However, the localisation and extent of these VLR and DBF are variable and raise the questions of their origin and natural evolution.

Aim of the study: To review the LLLySc with VLR and DBF and to try to define their origin and natural evolution

Material & Methods: Retrospective monocentric review of 135 LLLySc performed according to our standardized 3 phase protocol from 01/2021 to 1/02/2022.

Results: 24 patients had DBF (distal progression-DBF limited to the foot excluded: mean age = 62; range = 12-86: 7 men and 17 women: 5 had 2 exam: 6 had one history of previous cancer treatment-s and 9 of osteo-articular problem: BMI was higher than 30 in 14). 29 VLR-DBF were seen (at the level of the two limbs in 4, only right sided in 16 and only left sided in 5: most frequently at the level of the calf and/o the ankle) on WBS obtained after one hour of walking and their extent varied from very limited to extended from the foot to the inguinal area. The starting level was seen in 9 cases on the WBS after the 30 minutes in resting conditions, in 10 on the WBS after the tip-toeings (not done in 9 patients) and only after walking in 10.

Conclusion:

- In order to determine the starting point of one DBF, a dynamic and sequential approach should be performed
- Most of the DBF are rooted in (begin by) limited lympho-vascular lesions which raises the question of surgically correcting them and/or to limit manually their extension
- These DBF are 2 times more frequent at the level of the right LL than at the level of the left

THE INVESTIGATIONS OF THE DEEP LYMPHATIC SYSTEM OF THE LOWER LIMBS: SPECT-CT IS MANDATORY!

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Background: The lymphoscintigraphic investigations (LLySc) of the deep lymphatic system (DLyS) are now part of the management of lower limb edemas (LLE) and the visualisation of the popliteal lymph nodes (PopLN) is expected in these cases. However, these PopLN may be of deep and/or superficial lymphatic drainage.

Aim of the study: To review the clinical situations when these LLLySc of the DLyS were performed (after one deep injection of radiocolloids in the retro-achillear space), to analyze their results and to compare the contributions of the planar imagings and of the SPECT-CT imagings

Material & Methods: Retrospective monocentric review of 135 LLLySc performed according to our standardized 3 phase protocol from 01/2021 to 1/02/2022

Results: Among 26 patients (3 men and 23 women: mean age = 52.3 years: cases with Dermal BackFlow were excluded from the analysis) who had undergone one (bilateral) LLLySc of the DLyS, PopLN were seen at the level of 40 limbs (20 left and 20 right). On dynamic planar imaging performed with exercice (15 minutes long), PopLN could be related for the right limbs to medially and vertically running LV in 18 cases suggesting the presence of one deep lymphatic drainage (associated in 10 cases to internal saphenous LV) but only to the visualisation of one internal saphenous LV in 7. For the left limbs, the corresponding numbers were 17 (12) and/but 5. However, SPECT-CT could confirm the presence of one deep LV in the calf in only 19 cases and showed that in other cases these PopLN received their colloidal lymphatic activity from posterior superficial LV.

Conclusion: When the deep lymphatic system is investigated, spect-ct acquisition should be systematically carried in order to define whether deep and/or superficial lymphatic drainages are present, which determines the management of these patients.

THE POPLITEAL LYMPH NODES AND THE INVESTIGATIONS OF THE SUPERFICIAL LYMPHATIC SYSTEM OF THE LOWER LIMBS: SPECT-CT IS RECOMMENDED!

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Background: The visualisation of the popliteal lymph nodes (PopLN) in the framework of the lymphoscintigraphic investigations (LLySc) of the lower limb edemas (LLE) is common. However, these PopLN may be of deep and/or superficial lymphatic drainage.

Aim: to analyze and to compare the contributions of the planar imagings and of the SPECT-CT imagings

Material and methods: retrospective monocentric review of 135 LLLySc performed according to our standardized 3 phase protocol from 01/2021 to 1/02/2022

Results: 18 patients (4 men and 14 women: mean age = 62 years: cases with Dermal BackFlow were excluded from the analysis) had 23 PopLN (bilateral in 5: right-sided in 8 and left-sided in 5) seen on the planar LLySc (in one patient, the PopLN were faintly visualised on planar LLySc and could only be confirmed on SPECT-CT). In four cases, the presence of sural LN intercalated at the level of the calf could confirm that these PopLN were related to deep lympho-vascular drainage. In all but 3, normal internal saphenous Lymphatic vessel (LV) was seen and in 18 cases, the presence of one medially and vertically running LV suggested the presence of one deep lymphatic drainage. However, SPECT-CT could confirm the presence of one deep LV in the calf in only 11 cases and showed that these PopLN received their colloidal lymphatic activity from posterior superficial LV.

Conclusion:

- Popliteal LN could be definitely related to true deep lymphatic drainage in only half of the cases
- When popliteal LN are seen on planar imagings of LLLE, the realisation of spect-ct acquisition is recommended in order to define whether they are of deep and/or superficial lymphatic drainages, which determines the management of these patients

LYMPHOSCINTIGRAPHIC PATTERN IN CEAP 0S: ROLE OF DEEP LYMPHATIC CIRCULATION

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ESL Executive Committee Member, President of LymphoLab, Vice-President Italian College of PhleboLymphology

Topics: CEAP, Lymphoscintigraphy, Deep Lymphatics

Background: The diagnosis of chronic venous disease is essentially based on an accurate medical history and on the analysis of clinical manifestations. The CEAP classification is used to describe and classify chronic venous disease and includes not only clinical criteria but also aspects related to etiology, anatomy and pathophysiology.

The term CEAP 0 characterizes a picture devoid of visible or palpable signs of venous pathology. It can be asymptomatic or symptomatic (stage C0s) presenting pain, a sense of tension, skin irritation, muscle cramps and other symptoms attributable to venous dysfunction.

Many recent studies have confirmed the functional synergy between the lymphatic and venous systems. In particular, it was highlighted that, with the revision of the Starling principle, a large part of the drainage function of interstitial fluids is placed on the lymphatic system.

We know a lot about the role of the lymphatic circulation in CEAP 3 or in the non-healing pictures of advanced venous ulcerations, but what is the role of the lymph in the initial stages of venous disease?

Aim of the work: In our Center we deal purely with subjects with lymphatic problems. We wanted to investigate, through lymphoscintigraphic examination of the lower limbs, patients who came to our attention for a picture attributable to a CEAP 0s, to better delineate, even in these subjects, the role of lymphatic circulation.

Methods: 47 patients with clinical symptoms referable to CEAP 0s were investigated by lymphoscintigraphic examination and followed up with perometric scan. In all patients both superficial and deep lymphatic system evaluations were performed at three different Nuclear Medicine centers

Results: Patients undergoing lymphoscintigraphy demonstrated greater insufficiency of the deep lymphatic circulation. 63% of the patients presented deficiencies of this specific circulation to varying degrees.

In patients in whom a slowing of the deep circulation has often been demonstrated (76%), an edema localized to the fifth toe was also found semeiologically (an area often drained precisely through the deep lymphatic circulation) - Maccio's Sign

(It should be noted, however, that in all patients Stemmer's sign and objectivity were obviously negative for lymphedema and none showed an evolution towards a clear picture of lymphatic insufficiency)

Even if the patients did not report particular declining edema, we found that a simple medical sock with low Stiffness but of II kkl of compression (Circular knittign) allowed a reduction in the reported symptoms (C0s) in addition to a measurable volumetric reduction. Further data are needed, but it was still possible to observe an interesting predominance of subjects who presented functional alterations of the deep lymphatic circulation.

Conclusion: In conclusion we can say that functional relationships can be found between the lymphatic and venous circulation both physiologically and in the early pathological phases of the so-called chronic venous disease. The role of the deep lymphatic system and the role of shunts at the level of popliteal lymph node structures should be further investigated.

CAN LYMPHOSCINTIGRAPHY BE PART OF NUCLEAR MEDICINE OF THE THIRD MILLENNIUM?

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Introduction: The diagnostic and therapeutic approaches of many human pathologies have been substantially modified by the advent of extraordinary progress in the field of molecular and cellular biology. The validation of innovative imaging methods whose purpose is to visualize and quantify complex processes in vivo represent the future development in the diagnostic and therapeutic field of nuclear medicine. The future of Nuclear Medicine is therefore an effective use of radioactivity, which allows us to get more and more into the mechanisms that generate and maintain the disease and eliminate them or, if possible, to correct them [1].
As lymphoscintigraphy represents the "gold standard" for diagnosis of lymphedema, an important limitation is the lack of procedural standardization[6]. This is the first step toward certification of lymphoscintigraphy in the field of nuclear medicine of the third millennium.

Lymphedema: As well known lymphedema is a chronic and debilitating disease, often misdiagnosed, treated too late or not treated at all. Lymphatic injury, infection or congenital anomaly cause compromised lymphatic transport which causes lymphedema [2]. Lymphedema is found in both sexes, although women are investigated for this disease more often than men [3]. Lymphedema occurs at any age and two thirds of cases are unilateral. The distal part of the leg is initially affected, the proximal extension occurs later and even the feet are not spared[4].
In many countries lymphedema is still considered a "cosmetic" issue, rather than a chronic, degenerative and debilitating disease. The Italian Ministry of Health has adopted the guidelines for the National cure for lymphedema and related disorders, after a long period of study with internal and external experts [5].

Lymphoscintigrapy: The possibility offered to nuclear medicine in malfunction of the lymphatic vessels associated with the pathogenesis of many diseases, including lymphedema, fibrosis, inflammation and malignant tumors, allows it to be a primary protagonist in the development of the diagnosis and therapy of complex pathologies, including a series of rare diseases.
A further impetus to the development of nuclear medicine lies in the conception, design and construction of ever more efficient hybrid machines and/or design and synthesis of new hybrid radiopharmaceuticals, in order to provide more information with only one exam.
Imaging has improved significantly over the past 20 years and there are many tools and techniques currently in use to examine the lymphatic system that can be very useful for the diagnosis, staging and prognosis of diseases of the lymphatic vessels [3].
The advent of some of the newer techniques such as fluorescent imaging (usually with ICG) [7] and MRI and the newest non-contrasted MRI techniques show promise for the future with high spatial resolution.
To detect primary lymphedema, indocyanine green lymphography should be used first as a screening examination; when the results are positive, lymphoscintigraphy is useful to obtain further information.
At this point we asked ourselves in the super-technological era that we are experiencing, what the role of lymphoscintigraphy is in the entire diagnostic landscape.

Conclusion: About microsurgical LVA, Campisi et al. [8] conclude that previous lymphoscintigraphic diagnosing and subsequent lymphoscintigraphic follow up have a place in the surgical treatment of peripheral lymphoedema[9].
In patients with stage I extremity lymphedema, lymphoscintigraphy can predict the long-term response to physical therapy. Functional lymphatic changes detected by lymphoscintigraphy after external beam radiation therapy can predict the development of upper limb lymphedema[10].
Recently, lymphoscintigraphy, including single-photon emission computed tomography (SPECT)/computed tomography (CT) of the axillary region, has been employed to evaluate the impact of including, as target volumes in the radiation treatment plan, the lymph nodes involved in arm drainage that might affect lymphedema[11,12].
In pediatric patients, lymphoscintigraphy is the main diagnostic tool for clinically suspected congenital lymphatic abnormalities and in the case of edema of the limbs and genitals, chylothorax and chylous ascites[13].
Lymphoscintigraphy offers objective evidence to distinguish lymphatic pathology from nonlymphatic causes of extremity edema[14].
In conclusion Lymphoscintigraphy is generally the accepted technique of choice for diagnosing lymphedema because it is less invasive than other diagnostic methods but can still directly measure lymphatic dysfunction with relative ease[15].

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STUDY OF THE SUPERFICIAL AND DEEP LYMPHATIC COMPARTMENTS OF THE LIMBS: THE CONTRIBUTION OF LYMPHOSCINTIGRAPHY

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Introduction: Lymphoscintigraphy is the first choice examination for the diagnostic definition of edema, to confirm its lymphostatic nature, to identify the cause from obstacle or reflux, to evaluate the extent of the disease, the major impairment or less of the deep lymphatic circle than the superficial one, the drainage through the lymph node stations. It is therefore essential that the lympho-scintigraphic examination includes the study of both superficial and deep lymphatic circulation, through the appropriate injection of the tracer into the specific drainage locations of the two systems. The examination is not invasive, easily repeatable, also executable in neonatal age. It also allows to identify the IA stage of lymphostasis, which is still not clinically manifest, thus playing a fundamental role in the prevention of secondary lymphedema.

Aim: The aim of the work is to evaluate the involvement of superficial, deep or both circuits in limbs with and without clinical suspicion of lymphedema by lymphoscintigraphy.

Population and method: 254 patients (mean age 58 years, 70 males and 184 females) underwent lymphoscintigraphy of the superficial and deep circulation of the lower limbs (201) and upper limbs (53) by subcutaneous and intrafascial injection of 99mTc-nanocolloid . There were 167 primary lymphedema for the lower limbs (83%) and 7 in the upper limb group (13%). Total body images were recorded 20, 60 and possibly 120 minutes after the injection of the radiopharmaceutical. A quantitative multifactorial parameter (Transport Index) and a hepatic uptake index were calculated for each lymphatic circulation studied, both in the limbs with edema and in the clinically negative limbs. The oedematous limbs studied were 245 lower and 58 upper. Healthy limbs were 88 and 38 respectively.

Results: The study of the lower limbs shows between the affected limbs abnormalities of the superficial circle only in 16%, of the abnormal deep only in 19% and both pathological in 51%. Pathological TI was found in 46% of the limbs with no evidence of disease (only superficial 10%, only deep 24%, on both 12%).
The study of the upper limbs shows between the affected limbs abnormalities of the superficial circuit only in 12%, of the abnormal deep only in 2% and both pathological in 43%. Pathological TI was found in 31% of the limbs with no evidence of disease (only superficial 20%, only deep 9%, on both 2%).
The uptake index correlates neither with TI nor with the clinical stage of the disease.
A modest significance was observed between the interval between the appearance of the disease and the performance of lymphoscintigraphy and severity of IT in patients with superficial lymphedema of the upper limb. No significant correlations were found in all other groups.

Conclusions: The data derived from this work indicate an excellent sensitivity of lymphoscintigraphy in recognizing lymphoedema in the earlier stages of the disease. The scintigraphic study of both lymphatic circuits identifies a bilaterality of the disease in a high percentage of patients with apparently unilateral edema, allowing a more complete and effective therapeutic approach. Our case studies also suggest that most edemas recognize deep circuit involvement, while an anomaly in the superficial circle alone is rare. This data does not present substantial differences between upper and lower limbs and must be duly taken into account in the choice of the most advantageous microsurgical anastomotic procedure.

Topic: Different diagnostic approaches

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Thursday, June 16th, 2022

SESSION 2: PEDIATRIC LYMPHOLOGY

Chairs: Cestari M., Leone A., Moffatt C.

PEDIATRIC LYMPHEDEMA: GENERAL VIEW FROM DIAGNOSIS TO MANAGEMENT

BORMAN P.

Pinar Borman MD, PhD, Professor of PMR, Certified Lymphedema Specialist

Primary lymphedema; occurring as a non-syndromic inherited condition, or as a part of a syndromic disorder (Turner, Noonan, Prader-Willi, Klippel-Trenaunay, vascular malformations, is the most common type seen in children. In most cases edema is present from birth, but may also develop later in some cases. The awareness about pediatric lymphedema and early diagnosis is essential. The age of onset (congenital, pubertal), family history, location of swelling, associated conditions and dysmorphic features and underlying genetic causes are important in differential diagnosis of pediatric lymphedema. Diagnosis is based mainly on clinical presentation but physicians have to take complete anamnesis and extensive physical examination for coexisting systemic involvement and secondary causes. Physical examination should cover the assessment of skin changes (subcutaneous thickening, fibrosis, hyperkeratosis, papillomatosis), pitting edema, Stemmer sign and lymphangiectasia. If systemic involvement is present, ascites, pleural effusion and intestinal lymphangiectasia can occur. Lymphoscintigraphy is the gold standard for lymphatic system imaging in primary lymphedema but commonly it is not used in infant and toddlers. Abdominal US, venous Doppler US, MRI, x-ray, immune function markers, serum albumin and genetic testing can be helpful in differential diagnosis of pediatric lymphedema.

Early and accurate diagnosis is essential. There is no specific guideline for the management of pediatric lymphedema, consensus documents for adults can be adapted for children. Complex decongestive therapy comprising; skin care, MLD, bandaging, exercises, compression garments and self-management techniques, should be modified to the condition of each pediatric lymphedema patient according to the condition, site, degree of lymphedema and associated conditions. Bandages must not interfere with normal growth and restrict activities. Toes are not wrapped and delicate tissues of children require more padding. Education of families about skin care for prevention of skin changes and infection, self-management (MLD by families), compression and exercises are vital. Compression garments must be custom-madeare not recommended for children younger than 1 year-old. Compression garments should not exceed 20-30 mmHg of compression in children younger than 4 years of age. Psychosocial intervention for both children and their families should be taken into consideration .

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THE TRUE STORY OF THE REAL LYMPH I

From the publication "Angi y Cristobalito" (2nd ed.) and "La pequeña Linfa" (1st ed.)

BRAUN D., PAPENDIECK CRISTOBAL M.

Angiopediatria Fundation. Buenos Aires, Argentina.

Angi y Cristobalito is an illustrated clinical history about children with lymphedema. The purposes of the book are that children can identify themselves with these characters, recognizing what happens to them, understand in a didactic and simple way what a lymphedema is and also brin them closer to the health professionals involved in the treatment to facilitate communication and psychosocial integration.

From a fountain, the Little Lymph acquires an identity as a guide for young adults and parents about the Lymphatic System. Its step-pages are accompanied by the will and patience. The time witnesses the consequences that the secret code has for each of us. Friendship gives strength and waits for the great opportunity.

THE TRUE STORY OF THE REAL LYMPH II

"Cronik of de Lympedema in Pedriatics" (1st ed.)

PAPENDIECK CRISTOBAL M., BRAUN D.

Angiopediatria Fundation. Buenos Aires, Argentina.

The Little Lymph, Little for being pediatric, is a story that attempts to be a true story for a younger generation with interests , provoking the interlocutor to have a very sincere dialogue to know more about the mysterious reality.

It is not known what the term lymph means, nor lymphedema… going over what we know, very Little, but a lot to learn through a very sincere dialogue between two, the Little Lymph and me.

What is lymphedema? It is not a malformation. It is in some cases due to a malformation of the lymphatic system and others. Genetics, through molecular biology can explain some malformations as well as nematodes and mosquitoes, volcanic ash and other aspects ofcirculation, for example, the venous hypertension and more than 150 illnesses. All the lymphedemas are formed by water and proteins which relate in the interstitial space, which is not a space because it is full of wáter and something else. Is it an organ? Possibly the biggest of the anatomy, the organ with more water, and part of lymph, or all in time…

If everything is the same, why is it different?

Why is Chyle lymph and what is the volumen of lymph and why?

When leaves fall with the colours of time, they fall to stay as information has remained in time for more than 50 years. And we are on this surprising road, advancing. We hope that this simple information is worth knowing after at least reading it twice and with passion the great opportunity.

LYMPHOEDEMA IN DEVELOPMENT AGE: PRIMARY LYMPHOEDEMA TREATMENT

DI NICCOLO R.

Background: Lymphoedema is a particular condition of edema (from Ancient Greek "swelling") that increases the tissue consistence, caused by an high concentration of protein. It's a chronic, progressive and degenerative disease that leads to clinical complications and flare up. Primary grade, on genetic basis, are determined by an incomplete or abnormal development of the local lymphatic system. Secondary grade conditions set in after surgical lymph node dissection, radiotherapy or trauma.

Setting: Rehabilitation Institute "A. Quarto di Palo" (Andria).

Case report: 18 years old female patient.

Methods: Combined decongestive treatment, lymphotaping and constant use of the elastic restraint.

Results: Our team will explain patient's clinical evolution, rehabilitation and therapeutic aims.

Conclusion: We are going to demonstrate that patient's binomial compliance and multidisciplinary diagnostic and therapeutic approach, could stabilize or improve the clinical situation.

DIAGNOSIS AND THERAPY OF LYMPHEDEMA IN CHILDHOOD.

LEONE A.

Humanitas CCO Catania Italia.

The diagnostic and therapeutic pathway of lymphedema in paediatric age has peculiar characteristics in relation to the age of the patient, less cooperative, for the need for an early diagnosis, for the psychological aspects involving the child but also for the Family.

Clinical diagnosis must always differentiate primitive, hereditary and non-hereditary forms, from secondary forms, in pediatrics usually related with surgical acts or oncological pathologies, and from the syndrome forms, among the most frequent Prader-Willi, Turner, Nooan.

An accurate history and a painstaking visit, with the cooperation of the parents, is essential to frame from a clinical point of view the type of lymphedema and also to make differential diagnosis with other pathologies that I can cause edema.

Among the differential diagnoses need always take into account among the various possibilities in particular: chest or abdominal obstructive conditions, hypothyroidism, hypoalbuminemia, osteoarticular diseases, autoimmune diseases, angioedema Hereditary.

Instrumental diagnosis is based on the use of various methodologies: lymphoscintigraphy to get a clear picture of the anatomical and functional anomalies of the lymphatic system, for this survey should often be reassured parents about the safety of the examination, ultrasound for the study of skin and subcutaneous tissues, eco-color-doppler to identify vascular pathologies, abdominal ultrasound and abdomen MRI to detect pathological alterations of various kinds.

Lymphedema therapy involves doctor, physiotherapist, psychologist, nutritionist, and especially the small patient with the family; in children, the family plays a very important role in self-management of the disease.

Physical therapy includes manual lymphatic drainage, bandage, elastic brace, the use of velcro braces, such procedures should always be customized according to the age and clinical and family conditions of the small patient.

Medical therapy is based on the use of bromelin, safe even in paediatric age, coumarin and diosin in adolescence.

Surgical therapy, poorly indicated, better to wait a clear definition of the type of lymphedema before intervening.

Fundamental psychological support, for the family when the child is very small, for the patient in the stages of adolescence; For sports activity, often a source of psychological suffering due to limitations imposed by parents, a compromise must be found between the needs of the child and pathology.

Important to maintain the right body weight, avoiding obesity ensures better control of lymphedema.

FLT4/VEGFR3 MUTATIONS ARE ASSOCIATED WITH SEGMENTAL LYMPHATIC DYSFUNCTION AND INITIAL LYMPHATIC APLASIA IN PATIENTS WITH MILROY DISEASE

NINGFEI LIU, MINZHE GAO

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This study explored FLT4/VEGFR3 mutations and lymphatic defects in patients with Milroy disease (MD). Twenty-nine patients with lower limb lymphedemawere enrolled. Sixteen patients had a familial history of MD, while 14 patients exhibited sporadic MD. Clinical signs, FLT4/VEGFR3 mutations, indocyanine green (ICG) lymphography findings, and skin tissue immunohistochemical staining results were evaluated. Twenty-eight variants in FLT4/VEGFR3 were identified. Twelve variants have previously been reported; 16 comprised novel mutations. Of 28 variants, 26 were missense mutations; the remaining two comprised a splicing mutation and a nonframeshift mutation. Twenty-five variants were located in the intracellular protein tyrosine kinase domain; three variants were located in the extracellular immunoglobulin domain. Substantially delayed contrast-enhanced tortuous lymphatic vessels were visualized to the ankle or knee level in 15 of 23 patients who underwent ICG lymphography. No initial lymphatic vessels were visualized in skin specimens from four patients who did not exhibit lymphatic vessels during imaging analyses. No specific variant was identified in relation to the unique clinical phenotype. Segmental dysfunction of lymphatic vessels and initial lymphatic aplasia are present in MD patients with FLT4/VEGFR3 mutations.

This study is the first to identify a mutation in the immunoglobulin domain of FLT4/VEGFR3 in a patient with MD. It also shows that FLT4/VEGFR3 mutations are associated with two types of lymphatic defects in MD patients: one is a lymphatic collector dysfunction, which appears to be a common lymphatic pathology in MD, and the other is initial lymphatic aplasia in the skin. Moreover, lymphatic dysfunction was associated with mutations in both the immunoglobulin and protein kinase domains. Because of the heterogeneous lymphatic vessel pathology in MD, its treatment may require multiple therapeutic approaches.

DYSFUNCTION OF DERMAL INITIAL LYMPHATICS OF THE ARM AND UPPER BODY QUADRANT CAUSES CONGENITAL ARM LYMPHEDEMA

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Objective: The objective of this study was to explore the pathologic process underlying primary lymphedema.

Methods: Twenty-seven patients with unilateral congenital arm lymphedema who visited our clinic from January 1, 2014, to May 30, 2019, were enrolled. The patients' clinical signs and the findings of indocyanine green (ICG) lymphography, skin tissue immunohistochemical staining, and whole exome sequencing of tissue and blood were evaluated.

Results: Among the 27 patients, 11 were diagnosed with stage II and 16 were diagnosed with stage III lymphedema. No lymphatic vessels were visualized in the affected arm in 25 of 27 (93%) patients who underwent ICG lymphography; likewise, no lymphatics were found in the territories of axillary lymph node drainage in the trunk, irrespective of any anomalies of the axillary lymph nodes. In only two (7%) patients, an unclear lymphatic trunk gradually appeared in the dorsum of the affected hand. The number of initial lymphatics was increased in the skin specimens of all nine patients in whom lymphatics were not demonstrated by ICG lymphography. Among 14 tested patients, we found compound heterozygote variants in the PIEZO1 gene in only one (7%) patient. Two missense variants, c.4072C>T; p.Arg1358Cys and c.5033C>T; p. Ala1678Val, were identified and found to have been inherited from the father and mother, respectively. No other pathogenic or likely pathogenic variants of currently known lymphedema-related genes were identified in the remaining 13 patients. No genetic difference was found between the lymphedematous and nonedematous healthy skin tissue of the same person.

Conclusions: Segmental or regional dysfunction of the dermal initial lymphatics causes congenital arm lymphedema and may have implications for clinical treatment.

COMPLEX DECONGESTIVE THERAPY IN CHILDREN WITH LYMPHEDEMA

ROVNAYA A.

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Objective: Primary lymphedema (PL) in children is a challenge both for children and their parents. It can have its onset at any age and requires individualized approach in management and treatment. The common problem of PL in children apart from diagnostics, is treatment options, including self-management, that should be done mostly by parents.

Material-methods: a systematic literature - PubMed, Medline, Cochrane, and ALF, ILF, LE&RN, LSN resources were observed. Moreover, an analysis of our PL patients clinical cases (42), observed for 4 years was performed, to find the connection between severity of lymphedema symptoms and early/late start of CDT and the involvement of parents in the treatment process, and to understand what is the best way to adjust and supplement classic CDT for children of different ages.

Results: If CDT started more than 2 years after lymphedema onset, its results were not as good as when CDT started during the first year. The children, whose parents are performing CDT on regular basis (in addition to CDT in specialized clinic) and who comply to compression garments, show the best long-term results when compared to children who receive CDT in clinic only and wear compression garments. Orthopedic assessment and correction are crucial for muscle pump development and function if a child has orthopedic issues. Psychological support is essential in management of these children, as well it is very important that their parents get it too, because they are forming an environment in which a child is brought up and how he will see and accept himself and his condition.

Conclusion: After noticing PL onset, CDT should start as soon as possible. After 1st CDT phase in clinic, it should be done by parents on regular basis. Children with lymphedema should be observed by orthopedic specialists to choose the best option of orthopedic problems correction, if present. Both children with lymphedema and their parents need psychological support.

Thursday, June 16th, 2022

Workshop on Vascular Ulcers

Chairs: Pr Papendieck

LYMB BANDAGING FOR ACUTE LYMPHANGITIS AND LYMPHATICS ULCERS

MACCIÒ A., MD

ESL Executive Committee Member, President of LymphoLab, Vice-President Italian College of PhleboLymphology

Topics: Bandage, Wound care, Lymphangitis, Compression Therapy

Objective: Treatment of lymphangitis and lymphatic ulcers include two main approaches: prescribing antibiotics and bandaging. The choice of antibiotics depends on the severity of the symptoms or signs. The main goal of the bandaging is to reduce edema and improve peripheral lymphatic flow. There are various types of bandaging that differ in terms of execution and materials used.

Material & Methods: The multicomponent / multilayer lymphatic bandage or Maccio Lymphatic Bandage (LyMB) is used on patients with lymphangitis or lymphatic ulcer. The characteristics of the LyMB bandage are analyzed both by describing the different types of materials used and by evaluating the packaging technique that makes it a progressive and non-digressive pressure bandage.

The eight-point "lymphangitis score" is used to diagnose lymphangitis.

Results: The bandaging arises from the interaction between the specific knowledge of the materials used in each single layer (hysteresis and textile properties), the use of a specific bandaging technique to create a high static rigidity index (SII) and the indications given by patients. on their needs and requirements. Infact this type of bandaging results to have reached 99.8% of comfort felt up to 7 days from packaging

Conclusion: The correct execution of the bandaging and the knowledge of the materials to make it are the fundamental requirements to obtain an effective and well tolerated bandaging.

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Friday, June 17th, 2022

Session 3: Physical Therapy

Chairs: Hamadé A., Moneta G., Szuba A.

IMPACT OF ELASTIC DECOMPRESSION NEUROMUSCULAR TAPING METHODOLOGY MODULATING PHASIC MECHANICAL FORCES ON COLLECTING LYMPHATIC CONTRACTIONS

BLOW D.

NeuroMuscular Taping Institute, Rome. Italy

ABSTRACT: Amidst continuing incomprehension of taping techniques and substantial ambiguity of taping terminology for lymphedema, elastic compression aspects of taping resemble therapeutic effects of lymphatic bandage. Elastic decompression taping methodology is finely connected to physiological mechanisms of lymphatic drainage and control systems. Decompression, otherwise referred to eccentric (opposite to concentric) applications enhance the phasic contractile cycle providing a driving force to move lymphatic fluids. NMTaping support the primary mecanisms for lymphatic flow through pressure modification due to the skin-muscle-skeletal movement coordination dynamic. The oscillilation between a decompression taping action and local pathological lymphatic pressure is driven by active body mobility or by passive manual activation techniques, when the patient is not able to move, supporting intrinsic pumping of lymphangions. Elastic decompression hypothesis takes into consideration that interstitial and lymphatic fluid pressures are not static but continuously fluctuate with body and tissue movements. Continuous modification of pressure enhances fluid exchange mechanisms reducing possible tissue edema. 24 hour a day continual pressure modification is the underlying key and motivating force to this taping methodology

KINESIOTAPING IN LYMPHEDEMA

BORMAN P. MD, PHD, PROFESSOR OF PMR
Certified Kinesiotaping Practitioner (CKTP)

Kinesio-taping; as a lymphatic correction method, help to remove edema by directing the fluid towards a less congested lymphatic pathway and lymph nodes. The elasticity and lifting effects of kinesio-taping decrease the pressure on the superficial skin, open initial lymphatics and creates a massage effect during active motion. It also improves the transport of deeper lymphatics by allowing maximum contraction and relaxation of a muscle. Previous studies indicated better effects of kinesio-taping on decreasing lymphedema, when performed in addition to complex decongestive therapy. Fan type kinesio bandages are used in lymphedema anda re applied according to the lymphatic anatomy, anatomoses and site of lymphedema. Kinesiotaping can also be used as a therapeutic approach for scar formation in patients who have breast surgery or other surgeries. The duration of scar is not important as kinesiotaping improves all scars, independently from the duration of surgery. Also kinesiotaping is very helpful in patients with shoulder joint problems related with breast cancer surgery, as most of the physical therapy modalities (deep heat) used in shoulder problems, are contraindicated in cancer patients. Caution should be taken in patients with delicate skin and/or in children, in order to avoid side effects.

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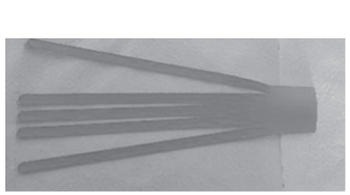









ANALYSIS OF DATA EXTRACTED FROM LINFOROLL® IN PATIENTS WITH SECONDARY LYMPHEDEMA IN THE PERIOD 2014 – 2018

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Linforoll® is a device created to perform lymphatic drainage and is able to show the physiotherapist in real time the body area treated, the pressure applied in mmHg, the number of steps to be performed on the work area, the speed of execution of the maneuver (cm / sec) and the energy transferred to the patient (Joule). These values are recorded and can be analyzed in order to obtain data that can guide subsequent treatments. Over the years the effectiveness of Linforoll® has been demonstrated within Complex Physical Therapy. This work aims to quantify, in patients with secondary post-surgical lymphedema, the value of the pressure exerted during the treatment sessions with respect to the volumetric and tonometric results obtained pre-post treatment, to quantify the treated areas (which, how often) with respect the results obtained in the context of the CPT and also to analyze the improvement percentages obtained with respect to the treatment modalities (pressure, number of therapies delivered in the different ranges of improvement compared to grading). Results: The pressure value range exercised was always over 100 mmHg. The pressure value exerted is often directly proportional to the increase in consistency (greater consistency corresponds to greater pressure exerted). The macro-zones of the lower limb (Thigh-Leg-Foot = 88.55%) and those of the upper limb (Arm - Forearm - Hand = 86.72%), were treated in a percentage way relevant compared to alternative pathways. All treatments were successful. Linforoll® also has an important action in lymph propulsion after lymphatic-venous microsurgery and after implantation of silicone tubes that bypass the lymphatic obstruction site. Linforoll is able to generate pressure gradients that allow the propulsion of the lymph towards the proximal end, where it can be absorbed.

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THE COMPREHENSIVE TREATMENT OF GIANT LYMPHOEDEMA

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Lymphoedema is one of the most complicated and difficult chronic problems to treat and cure for a long time and it has been a neglected medical condition.

I have been working in this field of medicine for nearly 35 years with my team. In our 35 years of experience we have tried various techniques and combinations to treat lymphoedema for a reasonable level of accepted success in these patients. Which includes the co operation of the patient, maintain the personal hygiene and the help of therapist to attain a soft , good skin quality and reduction and maintaining the reduction at that stage by pressure garments and therapeutic medication for preventing the cellulitis and secondary skin infection by periodic antibiotics and anti inflammatory and Lympho tonic drugs . Above all if the limb has lost its contours then we do different surgical techniques both physiological and reduction surgery to achieve a reasonably acceptable limbs and maintain it proper pressure garments . This comprehensive technique of treating lymphoedema with a TEAM of multiple speciality experts gives a better outcome in lymphoedema patients which will be explained in detail by a PowerPoint presentation in my oral presentations.

EFFECTIVENESS OF COMPLETE DECONGESTIVE THERAPY PHASE I (CDT) FOR THE TREATMENT OF LYMPHOEDEMA

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Introduction: Lymphedema is an abnormal accumulation of protein rich protein fluid in the interstitium which causes chronic inflammation and reactive fibrosis of the affected tissues.

Aim: Determine the reduction of circumferential measures of lymphedematous extremity after a daily 4 weeks complete decongestive therapy (CDT) program. Phase I consists of skin care, manual lymphatic drainage (MLD), bandage, exercises and education of the patient.

Method: We analyzed prospectively the therapeutic response of 105 patients with either primary or secondary lymphedema, who received treatment between 2017-2019. Patients' demographic and clinical characteristics were recorded. Excess volume (EV) and percent of excess volume (PEV) were measured at 4 times, during the treatment period; percent reduction of excess volume (PREV) was measured only once at last session.

Results: Among these 105 patients, 33 were with upper limb lymphedema and 72 with lower limb lymphedema. The majority of lymphedema was stage 2. A significantly reduction ($p<0.001$) between pre-treatment and post-treatment values of EV and PEV was observed. The median value of short-term reduction, estimated by PREV, was 66.5% and 71.5% in patients with upper and lower limb, respectively.

Conclusion: The management of lymphedema with CDT was successfully in all patients with almost the same results in both extremities. Prevention of the onset of lymphedema is of extreme importance.

THE PHYSIOTHERAPIST'S COMPETENCES WITH RESPECT TO THE " ITALIAN MINISTERIAL GUIDELINES ON LYMPHEDEMA AND OTHER PATHOLOGIES RELATED TO THE LYMPHATIC SYSTEM" AND THE " 2016 CONSENSUS DOCUMENT" OF THE INTERNATIONAL SOCIETY OF LYMPHOLOGY

GALLI T.

The Italian law 24/2017 sanctioned the definition of professional responsibility and protection of professionals when acting in reference to the guidelines and good practices.

Article 5 "Healthcare professionals, during the execution of health services with preventive, diagnostic, therapeutic, palliative, rehabilitation and forensic purposes, comply with the recommendations provided by the guidelines, except for the specificities of the concrete case ... and developed by public and private bodies and institutions as well as by scientific societies and technical-scientific associations of the health professions "

AIFI's Network Physiotherapy in Lymphology has aimed, among its objectives, at promoting the knowledge through recent scientific evidences and the Guidelines of National and International Scientific Societies.

The purpose of this study is to identify the physiotherapist's competences with respect to the "Italian Ministerial Guidelines on Lymphedema and other Pathologies related to the Lymphatic System" and the "Consensus Document" of the International Society of Lymphology.

Both documents consider CDP complex decongestive physiotherapy to be the golden therapeutic standard.

According to available evidence, the physical / rehabilitative treatment is the most appropriate way to achieve the best results in terms of clinical efficacy and maintenance of achieved results, as well as for preventing the natural progression of the disease.

We recommend to start a reflection on the approach to lymphedema, the conservative and surgical treatment, the early recognition, the patient empowerment, the self-management and the follow-up phase, in order to reach an integrated and interdisciplinary management strategy in an preventive perspective. In fact, in consideration of the evolutionary tendency of lymphedema, treatment should start as early as possible

PREVENTION (primary, secondary and tertiary) should guide the whole therapeutic process of the patient and represent the keyword of physiotherapy in all stages of lymphatic pathology: in secondary oncological lymphedema (from immediate post-operative phase in the case of lymph –adenectomy up to the palliative treatment phase) and in primary lymphedema (from diagnosis to stabilization).

The challenge to chronic diseases such as lymphoedema is a system challenge, which must go beyond the limits of the different professionals, promoting their integration and creating a model where the patient and his family become an active part of a personalized and shared care path.

To ensure adequate and high-quality assistance, it is necessary to encourage the establishment of integrated networks, in which hospital lymphological centers became a central and irreplaceable joint and activate surveillance and prevention programs. Moreover, these networks, through the definition of dedicated PDTAs, should allow the integration of all actors of primary, specialist outpatient and territorial assistance.

In relation to the rapid scientific evolution that is inevitably affecting and involving also our sector, the physiotherapist's skills, in addition to prevention, treatment, rehabilitation and functional assessment, are currently wider than what has already been established by our professional profile according to the DM 741/94 and by Law 251/2000.

In the lymphological context, as expressed in the previous points, **the physiotherapist assumes a function of a real glue (case manager) between the interprofessional team, the patient and his family, being an active part to prevent and reduce the fragmentation of assistance.**

THE PLACE OF GRADUATE DEGRESSIVE COMPRESSION SOCKS IN AMATEUR ATHLETICS AFFECTED BY LYMPHOEDEMA OF LOWER LIMBS

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Objective: The practice of some sports improves the quality of life in patients affected by lymphedema of lower limbs (LLL).The principal objective of this study is to determinate the place of graduate degressive compression socks (GDCS) in running in amateur athletics affected by LLL. The secondary objective is to evaluate the microcirculation of the feet in these athletics after exercise .

Methods: A monocentric observational study in 5 amateur athletics men aged from 28 to 50 years, affected by LLL (4 of left limb, the fifth of both leg) who practice running .All men are treated daily by GDCS. Duplex ultrasound, TcPO2, ankle-brachial-index (ABI), toe brachial-index (TBI) are normal. All men ran on treadmill in 3 exercises : 1 exercise without socks , he ran during the 2 and 3 exercise with GDCS on the leg affected by lymphedema (the 2 exercise with 20 mmHg, the 3 with 32 mmHg). We appreciated the effect of GDCS during exercise by questionnaire (pain, heaviness and mobility of leg affected by lymphedema).The calf circumference (CC), TcPO2, ABI and TBI are measured after each exercise.

Results: In terms of results, we have confirmed our objective of improving performance with GDCS during the exercise. All men reported reduction of pain and heaviness and performance of mobility with GDCS. CC is increased without socks and remains stable after the 2 and the 3 exercise. No significant change of TcPO2, ABI and TBI after exercise.

Conclusion: GDCS can improve the performance during exercise in amateur athletics affected by LLL. However a multicentric study with graduate degressive compression socks versus graduate progressive compression socks would be necessary.

THE EFFECTS OF TRUNK STABILIZATION EXERCISES ON BODY ENDURANCE AND PAIN IN LIPEDEMA WOMEN

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Objective: Lipedema is a chronic progressive adipose tissue disorder seen as a symmetrical enlargement of the hips and legs. In addition to aesthetic deformity, it is a disease in which women describe pain in the lower limbs. The aim of our study is to examine the effects of trunk stabilization exercises given to active joint range of motion exercises on body endurance and pain in lipedeme women. Methods: Thirty-two women with a mean age of 53.06 ± 7.92 years and diagnosed as lower and upper leg type of lipedema were included in the study. The cases were divided into two groups as experimental (n = 17) and control (n = 15) groups. All the cases included in the study were included in the exercise program, under the supervision of a physiotherapist 2 days a week for 5 weeks, and a home program for 5 days. The physiotherapy program of the control group consisted of active range of motion exercises, and the physiotherapy program of the experimental group consisted of body joint stabilization exercises in addition to the active range of motion exercises. After recording the demographic information of all cases, trunk endurance values and pain scores (Visual Analogue Scale) were evaluated. After the six-week treatment program, all evaluations were repeated. Results: As a result of our study, a statistically significant improvement in pain value was found in intragroup evaluation in both groups ($p < 0.05$); however, a significant difference was found only in the experimental group in the assessment of trunk endurance ($p < 0.05$). Looking at the evaluation between the groups; Experimental group was found to be statistically superior in body endurance values ($p < 0.05$). There was no significant difference between the two groups in pain scores ($p > 0.05$). Discussion: Active joint range of motion exercise program is effective in reducing pain in lower and upper leg type lipedematous women, while body stabilization exercises added to active joint range of motion exercises are a more effective method of improving body endurance.

OEDEMA OF LOWER LIMBS IN GREAT OBESE PATIENT: PROBLEMS TO SOLVE WAITING BARIATRIC SURGERY

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In recent years, it has become increasingly frequent to deal with the physical rehabilitation treatment of lymphedema or lymphedema with severe venous insufficiency on patients suffering, among other things, from severe obesity.

The experience in this regard involved taking care of 11 patients with lymphedema with concomitant venous insufficiency, 8 of whom with diffuse ulcerative lesions of various nature and morphology.These patients were all candidated to bariatric surgery, but with the necessity of a reduction of volume and often of solving of skin lesions before the surgical interventation. All the patients observed exceeded the weight of 200 kilograms with an age between 36 and 72 years. Among the observed patients, 6 had lipedemic component in association with lymphedema or phlebolymphedema. In all patients, the psychosocial aspect played a major role in all phases of physical rehabilitation treatment. 5 of them lived in poor hygienic condition and 3 presented a serial storage syndrome that promoted difficulties in resolution of sepsis of lesions. Another important aspect to consider is that in the 7 patients who lived alone, the difficulties of managing the treatment through elastic compression were added. The inability to bend down to remove the bandage and proceed to personal hygiene before repackaging it has led to important organizational and logistical problems. The bariatric surgery unit also requests the complete resolution of each skin lesion before intervening on the patient. In 2 patients, an important psychosomatic component was observed with psoriatic manifestations even in the injured areas or at risk of injury, leading the patient to repeatedly scratch the part in an uncontrolled manner, especially at night, worsening the clinical frame. In a patient with past following a deep venous thrombosys syndrome, the difficulty in resolving the lesions was fueled by a never diagnosed nickel allergy.

In a second patient with the same difficulties, the delay was related to the intake of a chemotherapy drug which, interrupted at the end of the cycle, showed to be the cause of superficial exudation and failure to resolve the lesion.

Once again the choice of materials and the 360 ° analysis of the psychosocial component is crucial in obtaining the results.

IS IT LOGICAL TO TREAT INTENSIVELY PATIENTS WITH LYMPHEDEMA IN AN OUTPATIENT STRUCTURE ?

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The authors bring their experience of intensive treatment of lymphedema from 1984 to 2019. The statistics are: 3180 treated in hospitalization (in patients): 703 primary (703 lower limbs, 5 upper limbs); 2472 secondary (2082 upper limb, 390 lower limbs).

The results are considered as good by the team and by the patients; of course after intensive treatment the patient is taken in charge by physiotherapist who provides lymphatic manual drainage, bandage, contention etc .. one or two times a week under the auspices of the personal physician of the patient (referent physician) .

The authors underline the very important link of the conception of safety attitude and they bring some complications: infections, cardiovascular, pneumological which demonstrate that this intensive treatment is impossible in an outpatient structure.

SELF-MANAGEMENT WITH COMPRESSION IN LYMPHOEDEMA

SCHINGALE F. J.

Lympho-Opt clinic

Abstract: Lymphoedema is a progressive, chronic disease as a result of primary or secondary damage to the Lymph system with consecutive increase and change in the interstitial tissue fluid, characterized by an alteration of tissues. The treatment of Lymphedema involves 2 phases and 5 columns, such as Phase of edema reduction and Phase of optimizing and maintenance with manual lymphatic drainage, skin care, Compression, exercises and self-management respectively.

The effects of compression: Compression therapy is recognized as an effective treatment in the management of venous and lymphatic disease since thousands of years. It is the main component of treatment. Due to the enhanced tissue pressure from outside by the compression, a reduction of the capillary fluid filtration follows as result, thereby less lymphatic water load results.

Objectives: Ideal goal of therapy is to normalize the lymphatic transport. Due to the chronic nature of lymphedema the therapeutic goal is to return the disease in the latent stage (limited transport capacity without lymphedema) or at least in the stage I and thereby achieve sustainable relief from the discomfort.

Methods: As in most of the countries all over the world we do not have enough experienced doctors and therapists that can do a qualified job on their patients, we have to educate our patients in compression, skin care and exercises. In the phase of reduction, we achieve the best results with a short- stretched bandaging. The stiffer the material the better the reduction. We need high working pressure which means that the bandage has to give a hard resistance to the muscle for better shifting of blood and lymph liquid to the heart. After about 5 hours the bandage will slip because the edema is reduced.

Therefore, the patient has to learn to bandage himself. Problem is, that nobody knows the pressure that is applied.

So we changed to Adjustable Velcro Wraps (AVW) for self-management, a stiff bandage with velcro closures and a BPS Guide what means: Built in Pressure System. The compression pressure is controlled with a card and two markers on each stripe of the device..

The same procedure with a normal inelastic multilayer bandage will take 3 times longer. In phase two we need compression hosiery to keep the reduced edema firm. During night time the patient can use the velcro device again to keep the result. In case that the edema starts to swell again, he goes back to the bandaging with velcro.

Results: Velcro device takes less time and the patient can adjust it after a few hours to control the necessary compression and there is a better reduction due to higher stiffness.

The patient is able to do the compression by himself and we know that we have the right compression for the corresponding disease.

Conclusion: Compression therapy and movement is the most important column of the treatment. Velcro devices are useful in both phases of treatment.

BENEFIT OF AN ADJUSTABLE COMPRESSION SYSTEM (COOLFLEX/COOLFLEX BOOT) FOR THE REDUCTION OF EDEMA (AKRÖ STUDY)- PRELIMINARY RESULTS

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Introduction: Adjustable compression systems are increasingly used to treat chronic edema due to their flexibility and ease of use. They enable the effective use of compression in contrast to bandaging, that requires the necessity for daily professional healthcare and the possibly of risking little improvement or even deterioration due to inappropriate bandaging technique. The Coolflex/Coolflex Boot compression system (Sigvaris) was designed to be donned and doffed with one hand, which makes it easier to operate for all patients, and can also be adjusted during the day, thus avoiding slipping and ensuring continuous adaptation.

Aim: This observational study aims to evaluate the effectiveness and ease of use of customizable compression systems (Coolflex/Coolflex Boot), as well as compliance and sustainability of compression therapy by patients.

Method: Patients with chronic leg edema (lymphedema, chronic venous edema, phlebolymphedema and other) and were recruited into this study. Exclusion criteria: PAD (ABI<0.5), phlegmasia, age <18 y.o. lack of consent. 77 consecutive patients with indications for compression therapy were included in 4 centers. Patients were examined and fitted for the Coolflex/Coolflex Boot compression system. The patients were assessed (measurements, questionnaire) after 3 days and six weeks.

Results: 69 patients completed the study. In 98.4% the wraps fit correctly after 12 weeks. 91% were able to putt the wrap independently. 94% worn the wrap ≥8 hours daily. 97% were satisfied wearing the wrap. Lymph-ICF UG Score improved significantly in all domains after 6 weeks. Leg volume improved in all patients.

Conslusions: Wraps allow independence of patients with chronic leg edema. The patients were satisfied and compliant with the new wrap system. Edema volume diminished in all patients after 6 weeks.

A HIGH FORCE 3 X 30 MIN COMPRESSION METHOD OF FOOT AND CALF REMOVES MOST EDEMA FLUID AND ENABLES IMMEDIATE USAGE HIGH COMPRESSION GARMENT

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Objectives: In advanced lymphedema of lower limbs stage III and IV bandaging under the routinely applied pressure of 30-50mmHg remains largely ineffective. This is caused by stiffness of skin and subcutaneous tissue due to fibrosis. Edema fluid is accumulating in multiple "lakes" what can be seen on the ICG lymphography. Evacuation of the stagnant fluid requires high external compression force and subsequent adjustment of non-stretch stockings to the decreased limb size.

Aim: To apply elastic rubber bandaging of foot and calf under the pressure of 90 to 125mmHg for 30 min on 3 consecutive days and evaluate fluid evacuation volume on plethysmography.

Material and methods: Twenty patients with lower limb lymphedema stage III of post-inflammatory type were included. Excluded were patients with acute inflammatory symptoms, venous thrombosis, profuse varices, obesity and cardiac insufficiency edema. A 10 cm wide rubber bandage was applied to foot and calf. The interface pressure on Picopress was 90 to 130 mmHg. Deep tissue tonometry, leg circumference and drop of interface pressure were measured. Strain gauge plethysmography was done before and after compression.

Results: Calf circumference decreased by 25±5%, deep tissue stiffness by 22±8%, interface pressure dropped to 45-70mmHg, plethysmography showed flat flow line indicating lack of mobile fluid.

Conclusions: High pressure 30 min leg compressions can remove excess edema fluid within 3 days and enable adjustment of pressure non-stretch stockings. This method is more effective at the beginning of therapy than the standard 30-50mmHg bandaging as it provides an immediate effect.

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Friday, June 17th, 2022

WORKSHOP ON PARTICULARITIES OF GENITAL LYMPHEDEMA: FROM DIAGNOSIS TO TREATMENT

Chairs: Michelini S.

WHAT DO WE KNOW ABOUT THE PATIENT PROFILE AND THE DIAGNOSTICS SPECIFICS

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Genital lymphedema is the chronic and progressive edematous condition of genital organs due to impairment of lymphatic circulation. It can be caused by primary or secondary lymphedema, and can affect both males and females, more common in males. The incidence or prevalence rates of genital lymphedema are not exactly known due to the lack of awareness, diagnostic criteria and delayed diagnosis. Primary genital lymphedema is the result of congenital dysplasia of lymph vessels and/ or lymph nodes, which may be present at birth or develop later in life with or without obvious cause. Treatment of gynecological, testicular, penile, intestinal or prostatic malignancies and melanomas existing around inguinal lymphatic areas, as well as trauma and infections may cause secondary genital lymphedema. The swelling may occur immediately after surgery or years later as with other forms of lymphedema. Various and different combinations of genital lymphedema may develop. Combined penile and scrotal swelling is more common in the males, while pubic areas and labias may be involved in females. The accumulation of lymph and the resultant inflammation stimulates adipose deposition and fibrosis which leads to papillomas and warty growths. Hyperkeratosis, lymphorrhea vesicular bleeding may also be seen. Cellulitis, wound and rarely hematuria, as complications of genital lymphedema may be life-threatening, if not treated early and aggressively.

An accurate diagnosis is important in providing appropriate management in this challenging condition which causes physical, emotional and social problems. Diagnosis of genital lymphedema depends on the detailed history, physical examination and differential diagnosis of other conditions In this presentation, genital lymphedema with different clinical presentations will be discussed in regard to etiology and characteristic findings.

NEW PERSPECTIVES IN GENITAL LYMPHEDEMA MANAGEMENT

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Introduction: Pelvic or genital lymphedema (PL/GL) is a chronic and challenging condition, that causes important discomfort and swelling altogether with emotional and social problems, leading to impaired quality of life for affected patients and families. In the absence of standard of care regarding treatment or evaluation criteria, it was decided to set up a non-comparative clinical exploratory study to assess the performance and safety profile of a new compression device in patients presenting PL/GL.

Methods: Between December 2020 and April 2021, thirty-two patients (17 men and 15 women) with PL/GL were enrolled in three reference centers for lymphedema in France. All patients included were advised to wear the MOBIDERM Intimate Short associated to the removable pad for 12 weeks (day and night). Primary endpoint was the patient's global impression of change (PGI-C) regarding quality of life related to PL/GL at 12 weeks. Secondary endpoints were quality of Life (LyQLI, EQ5D-5L), functional symptoms (swelling, tightness, discomfort, heaviness, pain), impact on sexual life, sleep quality, characteristics of lymphedema, compliance and safety.

Results: Twelve weeks after their inclusion, 78.6% of patients reported a global improvement of their condition related to PL/GL assessed by the PGI-C questionnaire. Patients also reported an improvement in quality of life with a specific impact in the psychosocial area, and a significant reduction of feeling of swelling, tightness, or discomfort. Physicians observed a clinically significant decrease of lymphedema severity, according to a decrease in Genital Lymphedema Score, a reduction of scrotum perimetry and an improvement in skin suppleness. No change in discomfort during sexual relations was reported during the 12-weeks study. Patients presented a relatively good compliance to the device with no sleep disturbance.

Conclusion: Use of MOBIDERM Intimate short shows benefit in patients with PL/GL. Nevertheless, these results should be confirmed in a well-designed RCT study.

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OF
lymphology
and related problems
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SESSION 4: COMPLICATIONS AND PREVENTION

Chairs: Amore M., Johansson K., Macciò A.

SIROLIMUS INDUCED LYMPHEDEMA

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Background: For some years now, the use of sirolimus has been increase to treat venous and lymphatics malformations, nevertheless this type of drugs is used as an immunosuppressant for solid-organ transplant recipients

Aim of this study: Evaluate the lymphedema induced by sirolimus.

Material & Methods: We evaluate 8 patients (N=8) who received sirolimus after kidney transplant (N=7) and liver transplant (N=1) for more than 10 years and recently they developed lymphedema after a minimal trauma, (N=6) unilateral lower limb and (N=2) combine unilateral lower and upper limb. We perform in (N=4) of this patients lymphocintigraphy of lower limbs, (N=1) Lymphocintigraphy of lower and upper limbs, (N=2) ICG Lymphography and (N=1) MR Lymphography.

Results: In all cases we observe hypoplasia and aplasia of lymph nodes with dermal backflow.

Conclusions: We need more evidence to know if that patients have a primary condition to develop lymphedema or the target of the rapamycin is focused on the involution of the lymphatic tissue. Maybe the dose and duration of the treatment have an impact factor.

EARLY INTERVENTION WITH COMPRESSION GARMENTS PREVENTS PROGRESSION IN MILD BREAST CANCER-RELATED ARM LYMPHEDEMA: A RANDOMIZED CONTROLLED TRIAL

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Background: Early diagnosis and compression treatment are important to prevent progression in breast cancer related arm lymphedema (BCRAL). However, in mild BCRAL, no progression can be expected in some patients, who may manage without compression. The aim was to investigate the proportion of women with mild BCRAL showing progression/no progression of lymphedema after treatment with or without compression garments, differences in changes of lymphedema relative volume (LRV), local tissue water and subjective symptoms during 6 months. Also, adherence to self-care was examined.

Material and Method: Seventy-five women diagnosed with mild BCRAL were randomized to a compression group (CG) or non-compression group (NCG). Both groups received self-care instructions, and the CG were treated with a standard compression garment (ccl 1). Women in the NCG who progressed in LRV≥2%, or exceeded 10% dropped out, and received appropriate treatment. The proportion showing progression/no progression of LRV, and changes in LRV was measured by Water Displacement Method. Changes in local tissue water were measured by Tissue Dielectric Constant, subjective symptoms by Visual Analogue Scale, and self-care by a questionnaire.

Results: A smaller proportion of LRV progression was found in the CG compared to the NCG at 1, 2 and 6 months follow-up ($p \leq 0.013$). At 6 months, 16% had progression of LRV in the CG, compared to 57% in the NCG, ($p=0.001$). Thus, 43% in the NCG showed no progression and could manage without compression. Also, CG had a larger reduction in LRV, at all time-points ($p \leq 0.005$), and in the highest TDC ratio, when same site followed, at 6 months ($p=0.025$). Subjective symptoms did not differ between the groups, except at 1 month, where the CG experienced more reduced tension ($p=0.008$). There were no differences in adherence to self-care.

Conclusion: Early treatment with compression garment can prevent progression in mild BCRAL.

Trial registration: ISRCT nr ISRCTN51918431

THE BENEFIT OF NO, IMMEDIATE OR DELAYED COMPRESSION GARMENTS TO PREVENT PROGRESSION IN MILD BREAST CANCER-RELATED ARM LYMPHEDEMA: A ONE-YEAR FOLLOW-UP.

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Introduction: A recent randomized controlled trial has shown that early treatment with compression garment for 6 months can prevent progression in mild breast cancer related arm lymphedema (BCRAL). Results in a longer perspective are however currently lacking. The **aim** of this study was to investigate i) the proportion of women with mild BCRAL showing progression/no progression of lymphedema after one-year follow-up, ii) differences in changes of lymphedema relative volume (LRV) measured by water displacement method (WDM), and in local tissue water by Tissue dielectric constant (TDC) ratio.

Material and Method: At end of RCT and start of follow-up 16 participants had no (NCG) compression and showed no increase of LRV, 17 had been treated with delayed compression garment (ccl 1)(DCG) when the threshold LRV increase ≥2% from diagnosis, and 32 participants had been treated with immediate compression garments (ICG) at diagnosis.

Results: At one year follow-up of NCG 14/30 (47%) participants still showed no progress of lymphedema, but 16/30 (53%) did. In the ICG compression was removed for one month after end of 6 months RCT and 24/30 (80%) showed no progression, but 6/30 (20%) did. In NGC 6 participants increase >10% and garment ccl2 was introduced, as well as for 3 participant in ICG.

Conclusion: It is beneficial for patients with early diagnosed BCRAL to start compression treatment immediate at diagnosis compared to delayed treatment after increase of lymphedema volume. However, some patients can manage without compression treatment at all.

LYMPHATIC ULCERS: HOW PHOTOBIOMODULATION HELP US?

MACCIÒ A., MD

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Background: Early diagnosis and compression treatment are important to prevent progression in breast cancer related arm lymphedema (BCRAL). HoLymphatic ulcer is a complication that can occur in advanced stages of lymphedema. As well as prescribing systemic antibiotics, the treatment includes the use of appropriate elastic bandages (zinc oxide or Manuka honey) to reduce edema, improve peripheral lymphatic flow and, in the case of Manuka, control bacterial overload. local. Furthermore, in recent times, resorting to photobiomodulation (PBM) is becoming an increasingly used clinical tool to induce the healing of wounds that are more resistant to traditional treatments

Aim of the work: Our center is specialized in treatment of lymphatic complications such as lymphangitis, ulcers and lymphorrhea. From our experience, proper bandaging is essential for the management of the ulcer. In addition, we wanted to test the new PBM method, one of the new frontiers of international wound care.

Material and Method: In this presentation, we will show you our specific experience about the use of a particular type of bandage LyMB combined with the PBM (LumiHeal™ system). The LumiHeal ™ system consists of two medical devices: a lamp, which emits blue light with a single peak wavelength between 440 and 460 nm, and a topical gel containing chromophores and urea peroxide. The molecules of the chromophore (not absorbed by the skin) absorb the light and convert it into dynamic fluorescent energy with a wavelength between 500 and 610nm capable of penetrating the affected tissues at various depths and promoting specific biological processes that should promote tissue healing

Results: In conclusion, our presentation underlines how positive results can be obtained through the use of appropriate bandages (for execution and use of the right materials) also supported by the use of PBM.

HIGH LEVEL OF LIPIDS RETAINED IN STAGNANT TISSUE FLUID/LYMPH MAY BE RESPONSIBLE FOR ADIPOCYTE HYPERTROPHY AND HYPERPLASIA IN LYMPHEDEMA

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Objectives: Advanced obstructive lymphedema is characterized as excess of interstitial fluid with proteins and overgrowth of fibroblasts and adipocytes. Albeit proliferation of fibroblasts and deposition of collagen has been documented, the mechanism of adipocyte hypertrophy and hyperplasia remains obscure. The question arises whether excess lipids in the tissue fluid/lymph in lymphedema is not a source of those accumulating in adipocytes. So far no studies in this direction have been reported.

Aim: To measure concentration of lipids in normal and lymphedema lower limbs tissue fluid/lymph and estimate how much can be retained under the condition of no lymph flow.

Methods: Thirty-five patients with lower limb lymphedema stage II-III served as tissue fluid/lymph donors. Edematous subcutaneous tissue was punctured and tissue fluid harvested. Concentration of the wide spectrum of lipids and apolipoproteins was measured.

Results: Total cholesterol 40±25 (serum 140±65), HDL 20±20 (42±12), LDL 10±12 (90±20), triglycerides 24±20 (32±18), APO1A 10±10 (140±80), APO B 6±2 (75±20), lipase 6±4 (22±4) mg/dL. Evidently high tissue fluid/level of cholesterol and triglycerides.

Conclusions: Present data confirm the previous ones that with the extracellular fluid volume of a lower limb 2.3L, the total mass of cholesterol is this compartment would be 460mg/dL and triglycerides 420mg/dL Retention of such evident mass in the tissue fluid/lymph under the condition of lymph stasis creates conditions for increased lipid absorption by adipocytes.

LIPOEDEMA: THE NEW HORIZON FROM INTERNATIONAL CONSENSUS DOCUMENT BY INTERNATIONAL LIPEDEMA ASSOCIATION (ILA)

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The speakers report about the International Consensus for Lipoedema, established by the International Lipedema Association (ILA). The Consensus Document reflects the international experts' shared view on the disease, following scrutiny of the available literature and at the same time taking into account many years of clinical practice with lipoedema patients. Just like the clinical complexity of lipoedema, ILA experts provided an interdisciplinary approach and as a result, a substantial change in prospective has taken place, not only in scientific understanding but also in the diagnosis and, above all, in the therapeutic approach to the disease. The ILA Consensus Document heart is that there is no scientific evidence to support the whole lipoedema dogma which has been previously published and propagated for decades. This lecture summarises the relevant findings of the consensus, emphasising the treatment of lipoedema now recommended.

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OF
lymphology
and related problems
VOLUME 33 . No. 82 . 2022
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Friday, June 17th, 2022

SESSION 5: LIPEDEMA

Chairs: Cornely M., Michelini S., Onorato A.

USA LIPEDEMA GUIDELINES

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[6]*Lymph Notes, San Francisco, CA, USA*
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[10]*Department of Physical Medicine and Rehabilitation, Vanderbilt, University Medical Center, Nashville, TN, USA*
[11]*Solace Massage and Mindfulness, San Diego, CA, USA*
[12]*Independent Contractor, Kansas City, KS, USA*
[13]*Olathe Health, Olathe, KS, USA*
[14]*Barth Syndrome Foundation, Larchmont, NY, USA*
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[16]*The Ohio State University Wexner Medical Center, Columbus, OH, USA*
[17]*Cleveland Clinic, Cleveland, OH, USA*
[18]*Fat Disorders Resource Society, Laurel, MD, USA*
[19]*Physical Medicine Institute, Orlando, FL, USA*
[20]*Campbell University School of Osteopathic Medicine, Buies Creek, NC, USA*
[21]*Yogamatrix Studio, Orlando, FL, USA*
[22]*Larson Plastic Surgery, Tucson, AZ, USA*

Background: Lipedema is a loose connective tissue disease predominantly in women identified by increased nodular and fibrotic adipose tissue on the buttocks, hips and limbs that develops at times of hormone, weight and shape change including puberty, pregnancy, and menopause. Lipedema tissue may be very painful and can severely impair mobility. Non-lipedema obesity, lymphedema, venous disease, and hypermobile joints are comorbidities. Lipedema tissue is difficult to reduce by diet, exercise, or bariatric surgery.

Methods: This paper is a consensus guideline on lipedema written by a US committee following the Delphi Method. Consensus statements are rated for strength using the GRADE system.

Results: Eighty-five consensus statements outline lipedema pathophysiology, and medical, surgical, vascular, and other therapeutic recommendations. Future research topics are suggested.

Conclusion: These guidelines improve the understanding of the loose connective tissue disease, lipedema, to advance our understanding towards early diagnosis, treatments, and ultimately a cure for affected individuals.

LIPEDEMA: USEFULNESS OF 3D ULTRASOUND DIAGNOSTICS

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Background: The diagnosis of lipoedema is based on clinical evaluation, but the high-resolution, non-invasive, repeatable and reliable ultrasound instrumental examination, relatively simple, even if operator dependent, is useful in the supra-fascial tissue study, using high-tech equipment with suitable high frequency linear probe.

Aim of the work: An evolution of standard ultrasound is high-quality three-dimensional imaging which, having a view on three planes of space, is interesting in the tissue structural study also for the possible processing of the image with appropriate software. For this reason, we have decided to apply this method in the tissue evaluation of lipoedema.

Material & Methods: In clinical stage I-II-III lipoedema, with the probe (17 MHz) placed directly on the skin, and longitudinal-transversal scans on constant markers along the limbs, the tissue ultrasound study includes the epidermis-dermis complex and the subcutaneous tissue.

Results: The 3D ultrasound picture has highlighted a normal ultrasound representation of the epidermis-dermis complex, an irregular, wavy or serrated profile of the junction dermo-hypodermis which, however, is not pathognomonic of lipoedema because it is also present in obesity-non-lipoedema and in lipo-hypertrophy.

Thickened hyperechogenic fibers, which connect the skin to the superficial fascia, and subcutaneous nodules, whose size varies in relation to the clinical stage, were highlighted.

The subcutaneous tissue has an increased thickness due to hypertrophy of the adipose lobules that appear adherent, but the disappearance of the interlobular connective septa and the fragmentation of the hyperechogenic fasciae, based on the evolution of the clinical picture, are not evident.

Conclusion: In this study in progress, 3D ultrasound diagnostics proved useful to evaluate the characteristics of suprafascial tissue in lipoedema and absolutely more detailed than standard ultrasound.

JOINT HYPERMOBILITY IS MORE FREQUENT IN LIPEDEMA THAN IN LYMPHEDEMA PATIENTS

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Background: Lipedema is characterized by a symmetric enlargement, fat accumulation, pain and frequent bruising in lower limbs. Recent studies point at a possible association between lipedema and joint hypermobility, but literature is still scarce.

Hypermobility is diagnosed though the Beighton scale when the score is greater than 3.

The estimated prevalence of hypermobility in general population is between 3-20%.

Aim of the work: We analyzed the prevalence of hypermobility in lipedema and lower limb lymphedema patients.

Material & Methods: We performed a prospective descriptive study of a sample of patients meeting lipedema criteria and a sample of lower limb lymphedema patients who attended the Lymphedema Unit. Data were analyzed using SPSS and mean, median and 95% confidence intervals were obtained. The correlation between the Beighton Score and the rest of variables was obtained through ANOVA and Chi-square tests. P-values <0.05 were considered statistically significant.

Results: We analyzed data from 81 patients with lipedema and 42 patients with lymphedema.

The median age was 52.1yrs (Range:18-80) for lymphedema patients and 44.6yrs (Range: 18-68). Mean BMI was 25.4 (95% CI:23.9-26.8) in patients with lymphedema and 28.5 (95% CI: 27.1-29.8) in lipedema. Mean waist-height ratio was 0.50 (95% CI: 0.47-0.53) in lymphedema and 0.50 (95% CI: 0.48-0.52) in lipedema.

Mean Beighton Score was 4.06 (95% CI: 3.52-4.06) in patients with lipedema and 2.60 (95% CI: 1.92-3.27) in patients with lymphedema.

While 45.7% of all patients with lipedema were hypermobile according to the Beighton score, this was true for only 26.2% of all patients with lymphedema, which is nearly a two-fold increase in prevalence between both entities. More interestingly, the knee extension test of the Beighton score was bilaterally positive in 18.5% of the patients with lipedema and 4.8% of the patients with lymphedema.

Conclusion: Lipedema may have other clinical manifestations besides those widely known. Joint hypermobility seems to be more prevalent among these patients as compared to patients suffering from lymphedema, even though limb volume is increased in both. The largest difference was seen in the knee extension test, where we obtained nearly a four-fold increase between patients with lipedema and patients with lymphedema.

MEASUREMENT AND ASSESSMENT OF LIPEDEMA:

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Lipedema is defined as a painful disproportionate fat distribution disorder; it affects nearly exclusively women. An increasing scientific interest is attributed to it, in particular with regard to pathophysiology and surgical treatment. From the point of view of its measurement and evaluation, there are still several aspects that deserve further investigation, to offer the clinician various possibilities aimed to correctly identify the condition, to quantify and monitor the extent and relevance of symptoms and signs, to distinguish associated pathologies, as obesity, venous or lymphatic circulation disorders or endocrine pathology.

A first aspect is the accurate collection of the anamnesis, which takes into account all the factors (especially the modifiable ones) that favor or aggravate lipedema. The physical examination is another relevant step, due to the possibility of identifying all the elements that can confirm or not the presence of the pathology, as well as allowing its staging and classification.

A further aspect is represented by clinimetrics, with the possibility of using different instrumental studies.

The approach is multimodal and involves the possibility of collecting, if possible in an objective way, the following data: body mass (with BMI) and waist-hip-ratio, volume of the different segments (with indirect method -cirtometry- or direct -water volumetry-); body morphology (recorded with photographic apparatus, with body scanner), entity of body compartments (with measure of impedance, with DEXA); quality of affected tissues (by ultrasound, possibly with an elastographic method; tonometry); coexistence of lymphedema (with ultrasound, with lymphoscintigraphy); extent of pain (with dedicated scales, with algometers), psychological status and quality of life.

The choice of the methods used may depend on the purpose that the team can have: simple patient diagnosis and monitoring or even research purposes, in particular to evaluate the impact that the various therapies currently proposed can have on lipedema as well as on the psychological sphere of the patient.

Some of these approaches can be misleading, if not well understood by the clinician.

There is no doubt that as in other fields of medicine, where the diagnostic definition can be complex, even in the study of lipedema the coupling of multiple diagnostic modalities will lead to a better understanding of the pathology and a better choice of the appropriate therapeutic approach.

WHAT ARE THE MOST IMPORTANT FEATURES OF LIPEDEMA?:CLINICAL RESULTS FROM A PROSPECTIVE COHORT STUDY.

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Lipedema is characterized by bilateral enlargement of the lower limbs (LL) due to abnormal depositions of subcutaneous fat, pain and bruising. Diagnostic criteria are not stated, and its tendency to progress is controversial.
Aim: To describe signs and symptoms of lipedema patients and the change of LL volume at long-term.
Design: Prospective cohort study of lipedema patients.
Female patients were included if they present with bilateral LL enlargement and at least 3 symptoms of lipedema: spontaneous pain/tenderness, easy bruising, family history, absence of LL injuries, absent Stemmer sign, symmetrical LL involvement, no swollen feet; and consent to participate.
The endpoint was the prevalence of symptoms and signs present in lipedema.
The type, stage of lipedema, type and intensity of pain and orthopedic alterations were also assessed.
To describe the evolution, we calculated the Percentage of Volume Change (PVC) during follow-up. Progression of disease was defined when PVC>10% from baseline; Improvement when PVC<-10% and Stability when PVC:-10 to 10%**Results:** We analyzed data from 81 patients with lipedema and 42 patients with lymphedema.
The median age was 52.1yrs (Range:18-80) for lymphedema patients and 44.6yrs (Range: 18-68). Mean BMI was 25.4 (95% CI:23.9-26.8) in patients with lymphedema and 28.5 (95% CI: 27.1-29.8) in lipedema. Mean waist-height ratio was 0.50 (95% CI: 0.47-0.53) in lymphedema and 0.50 (95% CI: 0.48-0.52) in lipedema.
Mean Beighton Score was 4.06 (95% CI: 3.52-4.06) in patients with lipedema and 2.60 (95% CI: 1.92-3.27) in patients with lymphedema.
While 45.7% of all patients with lipedema were hypermobile according to the Beighton score, this was true for only 26.2% of all patients with lymphedema, which is nearly a two-fold increase in prevalence between both entities. More interestingly, the knee extension test of the Beighton score was bilaterally positive in 18.5% of the patients with lipedema and 4.8% of the patients with lymphedema.
Results: From September-2012 to December-2019, 138 patients were included. The median of age was 47.6yrs (range:18-80), and the time-elapsed-until-diagnosis was 25.5yrs (+15.8).
The most frequent type of lipedema was Type 3, from the ankles to the hips, in 2/3 of the patients. And most were in stage I and II.
In our sample, 23.2% of the patients have normal BMI, 31.2% are overweighed, 37.7% are obese and 8% have morbid obesity.
Considering the recommendation to use waist-to-height-ratio for lipedema patients, as it is a better indicator of body fat distribution, 41.3% were slim or healthy; 19.6% are overweighed and 36.1% are obese or morbid obese.
The most frequent features of lipedema were the symmetrical involvement of LL, the unaffected feet, pain and easy bruising.
Family history was positive in 85%.
Clinical orthopedic examination revealed cavus feet and valgus knees in 1/3 of the patients. The XRay study was performed in 63 patients, and knee osteoarthritis was diagnosed in different stages of the disease. Hypermobility was diagnosed in 45.7%.
The greater the age, the more advance stage of lipedema: stage I (mean age: 38.0 CI95%: 34.7-41.3), stage II (mean age: 48.8 CI95%: 45.0-52.6), stage III (mean age: 58.1 CI95%: 53.6-62.6), stage IV (mean age: 56.0 CI95%: 49.5-62.5) (p<0.0001).
The Waist-to-hip Index increases with the age (β: 0.003; p<0.0001).
In more advanced stages of lipedema, Stemmer' Sign was more frequently positive: stage I (3.8%), stage II (14.6%), stage III (19.4%), stage IV (57.1%) (p<0.0001).
The greater the BMI the more advanced stage of lipedema, Stage I: (mean of BMI; CI95%) 25.5 (24.4-26.6); Stage II: 30.3 (28.7-32.0); Stage III: 34.5 (32.5-36.5); Stage IV: 38.8 (34.1-43.6); p<0.001.
From 138 lipedema patients, follow-up was performed in 69 patients, at a median of 3.8yrs after baseline.
Compliance to Maintenance therapies was MLD 8.7%; Intermittent Pneumatic Compression 5.8%, Bandages 5.8%; Compression garments 69.6% and exercise 52.2%.
Patients that were compliant to compression garments wore them 11.4 hours/day (95%CI:10.6-12.2), 6.0 days/week (95%CI:5.7-6.4), half wore plain tissue and most of them wore pantyhose (76%) and ccl2 (96%).
The mean PVC: 4.3% (95%CI:1.7-6.8). Stability of the volume was observed in 61.2% of the patients, progression in 32.8% and improvement in 6.0%.
A significant positive association was observed between PVC and weight change. The more the weight increases, the more lipedema progresses (β:0.599; p<0.0001).
The age was related to less progression of lipedema (β:-0.280; p=0.022).
No relation was observed with chronicity; stage of lipedema nor the presence of lymphoscintigraphic alterations.
No associations were observed with MLD, IPC, Bandages, exercise or compression garments compliance.
Conclusion: Type 3 is the most frequent type of lipedema.
Obesity is a frequent comorbid condition but is not so prevalent in our sample than in other studies that consider normal BMI as a rarity.
The most frequent features of lipedema were the symmetrical involvement of LL, the unaffected feet, the pain and easy bruising. These could be considered as Major criteria for diagnosis.
Clinical orthopedic examination is important. Knee pain is frequent, and can be impairing the ability to comply with the exercise recommendation.
We observed that the more advanced stage of lipedema was more frequent among older patients, and this could suggest the progressive characteristic of lipedema. This is controversial as many authors state that lipedema is not progressive along the time.
In the follow-up study, Lipedema was stable in 2/3 of the patients. Progression was related to weight increase. Further research is warranted to detect the patients that are going to progress despite treatment.

LIPOHYPERPLASIA DOLOROSA OR LIPOEDEMA. WHICH TERM IS BETTER FOR THIS CONDITION?

PROF. CORNELY M.

"The basic difficulty in lipedema is the deposition of an unusual amount of fat beneath the skin". This sentence by Allen and Hines, published in 1940, directs the attention directly to the tissue that is in the foreground in the description of the disease called "lipedema" by the first authors, today - better studied - called lipohyperplasia dolorosa (LiDo). Fatty tissue gives shape to the body, as does oedema. The two internal medicine authors saw subcutaneous fat distribution on the legs and orthostatic oedema. This clinical observation led to the misnomer "lipedema".

The disease "lipedema" is increasingly better understood. In particular, oedema is not significant in the fat distribution disorder of women's arms and legs. This and other scientific findings are "work in progress" with the aim of renaming the disease. A proper name is "Lipohyperplasia dolorosa" (LiDo). In lipohyperplasia dolorosa, the increase in volume is genetically fixed, but the pain is dynamically progressive. LiDo describes the congenital genetic symmetrical pressure-painful fat distribution disorder of the legs and arms, taking into account the known histological adipocytic variant. LiDo also occurs on the arms in up to 90% of patients. This distribution pattern had not been noticed by the first describers.

There is no indentable oedema, so that the term lipohyperplasia dolorosa (LiDo) rightly omits the suffix "oedema". Passive orthostatic oedema may occasionally occur in the legs, but never in the arms, in addition to the typical fat distribution disorder. Today it is possible to describe more precisely than 80 years ago what comprises and characterises this clinical picture in women and how patients can be helped.

LiDo must be distinguished from other symmetrical, painless fat distribution disorders of the arms and legs prima vista and, after palpatory examination, especially from coincident obesity.

Obesity is never comorbid, but often coincident with LiDo. Physical activity and dietary changes can reduce obesity but cannot eliminate the disproportionate fat tissue increase in the extremities caused exclusively by LiDo. In LiDo patients coincident with obesity, there is no effect of gastric surgery on the obligatory painfulness of LiDo.

There are both conservative and surgical treatment options for lipohyperplasia dolorosa. One established procedure since 1997 is surgical treatment by Lymphological Liposculpture™ . The most important result of consistent surgical treatment of lipohyperplasia dolorosa is the influence on the quality of life: "It ruined her life" becomes "It improves her life".

COMPARISON OF THE EFFECTIVENESS OF LOW-CARBOHYDRATE, HIGH-FAT DIET WITH MODERATE-CARBOHYDRATE AND FAT DIET IN REDUCING BODY WEIGHT AND BODY COMPOSITION PARAMETERS IN PATIENTS WITH LIPEDEMA

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Objective: The aim of this study was to compare the effectiveness of two diets: low carbohydrate high fat (LCHF) and medium fat medium carbohydrate (MFMC) in reducing body weight, body fat and lower limb circumference in patients with lipedema.

Material & Methods: The study group consisted of 91 women (mean age: 44.0 ± 13.5 years) with lipedema. The participants were divided into two groups following two interventional diets for 16 weeks: low-carb high fat diet (LCHF; <10% energy from carbohydrates; >70% energy from fat) and medium-fat and carb diet (MFMC; <40% energy from fat and carbohydrates, equally), which were applied to indicated caloric restrictions. Anthropometric measurements were made at the baseline and after 16 weeks.

Results: Both LCHF and MFMC diets contributed to significant reductions in body composition parameters and body circumferences in patients with lipedema. The difference of body weight after dietary intervention was: -8.2 ± 4.1 kg vs -2.1 ± 1.0 kg for LCHF and MFMC diet, respectively (p<0.0001). The percentage body fat decreased by -4.2 ± 2.1 % in LCHF group, and by -0.9 ± 0.5 % in MFMC diet (p<0.0001). Both diets contributed to decrease of visceral fat level: -2.6 ± 1.3 in LCHF diet, and -0.5 ± 0.3 in MFMC diet (p<0.0001). LCHF diet contributed to greater reduction of waist and hips circumferences than MFMC diet. The circumferences of thighs and calves decreased significantly in both groups, but more effective in LCHF group.

Conclusions: The LCHF diet was more effective than MFMC in reducing body weight, body fat and lower limb circumferences in patients with lipedema.

ONSET OF DERCUM'S DISEASE (ADIPOSA DOLOROSA) FOLLOWING C0VID -19 INFECTION

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Objective: Dercum's disease is a rare disorder of unknown etiology that presents with painful subcutaneous fatty growth of various sizes. We present 4 cases of Dercum's disease development following Covid-19 infection.

Methods: Case 1 is a 31-year-old woman with stage I lipedema with new onset of multiple subcutaneous painful nodules at different areas of her body 3 months following Covid- 19 infection. Case 2 is a 66-year-old diabetic woman with history of left upper extremity swelling, painful nodules in the left breast, left arm and abdomen 5 months following Covid-19 infection. Case 3 is a 56 years old secretary complaining of painful gluteal area and upper thighs, inability to sit for any length of time, fatigue, foggy brain and difficulty to complete her work. Her laboratory test results were negative. She was referred to a psychiatrist. The Ultrasound study and clinical examination confirmed Dercum's nodules and Lipedema. Case 4. is an athletic 36 years old woman with bilateral, painful axillary mass after a prolonged Covid-19 infection. The clinical evaluation and Ultrasound study confirmed Dercum's Nodules and Lipedema.

Results: This study demonstrates the importance of early and correct diagnosis of Dercum's disease and Lipedema confirmed with ultrasound study visualizing multiple "blush–like" hyperechogenic subcutaneous fatty nodules of different sizes.

Conclusions: Ultrasound study objectively measure the severity of the disease. Early diagnosis and early treatment may prevent further progression of the Dercum's disease.

ESL CONSENSUS DOCUMENT ON LIPEDEMA: PROPOSAL FOR DISCUSSION

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Lipedema is a group of undefined diseases that recognize in a pathological growth of adipose tissue in certain body regions still to be investigated, understood and defined from a nosological point of view.
The clinical framework common to the different syndromic aspects is determined by a symmetrical accumulation of subcutaneous fat, chronically progressive, accompanied with orthostatic edema, which almost exclusively affects female sex. In most cases the affected subjects have a normal body mass index and, despite this, many clinics still today believe that the disease is very linked to some epigenetic aspects, especially wrong diets with consequent incorrect address of patients to bariatric surgery with further damage. The etiology, to date, is still not fully defined. In 2020 our team discovered the first gene that, if mutated, is responsible for the phenotype: alpha-ketoreductase c1, an enzyme involved in the metabolism of progesterone. The role of hormones is also undoubtedly important; this is demonstrated by the almost exclusive involvement of the female sex, the onset at puberty and the accentuation of the clinical picture during pregnancy and at the time of menopause.
The relationship with obesity is not clear. it is certainly not systematic. In our experience, many subjects have a body weight even lower than normal, because those affected avoid part of the daily food intake, with behaviors bordering on anorexia, to avoid, by taking in calories, an increase in tissue volume. It is clear from a conspicuous Literature that diets low in calories do not have the desired effect on the areas of the body affected by lipedema, as does physical exercise, to which other adipose deposits of the same organism respond. Conversely. The weight increase is primarily poured into the affected areas.
The common clinical aspects of the disease are represented by familiarity. The distribution of fat occurs essentially in the buttocks, thighs and legs for the lower limbs, in the arms and forearms for the upper limbs. There is a clear demarcation between affected tissue and healthy tissue at the ankle and wrist with savings of the feet and hands. The development of the diseased is simultaneous, bilateral and symmetrical. Impressive oedema is typical of the advanced stages of the disease. there is a poor response to exercise and diuretics. Pain is present in over eighty percent of subjects and is accentuated with acupressure. Spontaneous hematomas in the affected limbs are frequent.
There are four clinical stages: in the first there is evidence of edema with an unchanged tissue consistency; in the second beginning, on palpation, fat granules are appreciated which are distinguished from the rest of the edematous tissue; in the third stage there is marked deformation of limbs and in the third severe stage (also called fourth stage) the deformation of the limbs is monstrous and imposes great functional limitations. In relation to the localization of the abnormal adipose tissue, five types have been conventionally described: localization only to the buttocks, localization that extends to the thighs, which also affects the legs, which affects the upper limbs or only the legs. In reality, in daily clinical practice, it is possible to observe as many as 9 different types of localization that can vary affect only the localization of the lower limbs, in the various locations, or, at the same time, both the lower and upper limbs
Lipedema is accompanied by local circulatory abnormalities such as microangiopathy with consequent vascular fragility so that lipedema patients are predisposed to spontaneous bruising.
Severe localized pain is reported when the medial below the knee joint area (at the fibula head level) is pinched deeply. it is a pathognomonic sign of lipedema which is present from the early stages of lipedema. hypodermal adipose tissue ultrasound show normal dermal thickness and an increase of subcutaneous tissue thickness and subcutaneous echogenicity due to the hyperplasia/hypertrofia of the adipose cells and the presence of fibrosis. Another ultrasonographic typical aspect is the disappearance of interadiposis septa. Echography is also useful for the differentiation between lipedema and lymphedema, this last characterized by dermal and subcoutaneous thickness increased and subcoutaneous anechogenicity due to the interstitial fluid.
Another relevant aspect in ultrasoud examination of lipedema is that the compression of the skin shows a reduction in the sub-fascial thickness while the supra-fascial thickness remains unchanged. this confirms the presence of adipose tissue cells that cannot be confused with interstitial fluid typical of lymphedema ultrasonographic pattern.

In patients with lipedema lymphatic system of the lower limbs, unlike patients with primary or secondary lymphedema, is normally developed and functioning. Lymphoscintigraphic examination performed in these subjects only in the third stage shows signs of transport deficits related to extrinsic compression by the adipose mass of the lymphatic vessels themselves. In these cases it is possible to highlight dermal back flow, or, more commonly, abnormal lymph node stops along the course of the limbs.

In advanced clinical stage of lipedema the reduction of motion and the extrinsic obstruction of lymphatic vessels can develops a more or less important lymph-stasis to treat in appropriate way

Psychosocial distress and associated mental health conditions such as depression, anxiety, body dissatisfaction, and eating disorders may contribute to not only a decrease in quality of life but also to further progression of the disease.

There are other comorbidities associated to lipedema. In our experience, according to several recent clinical studies, we found cases of insulin resistance, Hashimoto thyroiditis, low levels of vitamin D, articular inflammation, hypermobility, endometriosis, alterations in the menstrual cycle, with different percentage of recurrence.

Hypermobility is a condition reported in association with lipedema. ~ 15-20 % of women with lipedema had a positive Beighton score (≥ 5). Joint hypermobility could be associated to Ehlers Danlos syndrome hypermobility. Actually they are two different therapeutical approaches for this pathology: The first is conservative, the second one surgical. But, still today, there are no guidelines that firmly indicate when and what to choose. There are still few clinical trials conducted on the management of this disease sometimes with discordant results. aspects that must certainly be taken into consideration with regard to the type of therapeutic choice to be made, such as the degree of pain of patient, disability, presence of comorbidities, any compliance with conservative treatment, economic and non-financial resources. Patient's will must be taken in consideration at the same time.

Last year the USA guidelines on lipedema were published. The document, in particular underlines above all the hereditary and genetic nature of the disease, confirms that traditional low-calories diets, physical exercises and bariatric surgery do not solve the clinical problem, confirm the proscription of diuretics for therapy and the positive use of metformin, above all in subjects with insulin resistance, confirm the need of vitamin D and the importance of the combined physical treatment both in the containment of the disease and in the regression of the main symptoms (first of all the pain). The team of health workers must be culturally and practically well prepared. The document conclude with that there are two surgical techniques very effective for lipedema surgical treatment (among the many currently used) to maintain the clinical results along the time: the WAL (water assisted liposution) and the PAL (power assisted liposuction).

Lipedema is a chronic painful disease. So a correct diagnosis is required in order to not incur in wrong and not resolutive treatments. Beside the aesthetical aspect evaluate the functional discomfort and the sense of frustration in front of expensive and useless therapies. Early diagnosis is necessary to prevent the clinical evolution of the disease.

The different syndromic frameworks confirm that this is a cluster of adipose tissue diseases yet to frame in their nature having in common the presence of abnormal cells, the behavior of which needs to be able to make a special appropriate therapy.

In conclusion, lipedema is a rare genetic disease that is affected by both the genetic predisposition and some epigenetic aspects that must still be in-depth.

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MOLECULAR PATHWAYS INVOLVED IN PRIMARY LYMPHEDEMA

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Lymphedema is a chronic inflammatory disorder caused by ineffective fluid uptake by the lymphatic system and limited lymph flow with effects mainly on the lower limbs. Lymphedema is either primary when caused by genetic mutations, or secondary when it follows injury, infection, or surgery. In this study, we aim at assessing to which extent the current genetic tests detect genetic variants of lymphedema, and at identifying the major molecular pathways that underlie this rather unknown disease. We recruited 147 individuals with clinical diagnosis of primary lymphedema and used established genetic tests in their blood or saliva specimen. While only 11 of these were positive, other probands were either negative (63) or inconclusive (73). The low efficacy of such tests called for greater insight into the underlying mechanisms to increase accuracy. To this purpose, we built a molecular pathways diagram based on a literature analysis (OMIM, Kegg, PubMed and Scopus) of candidate and diagnostic genes. The PI3K/AKT and the RAS/MAPK pathways emerged as primary candidates as responsible for the lymphedema diagnosis, while the Rho/ROCK pathway appeared less critical. At present, only few studies have focused into the molecular pathways underlying lymphedema and the validity of genetic tests for its diagnosis remains limited. The results of this study suggest the most important pathways involved in the pathogenesis of lymphedema and outline the most promising diagnostic and candidate genes to diagnose this disease.

NATIONAL REGISTER OF RARE DISEASES, CASES WITH RARE DISEASES ATTENTION CODE RGG020 FROM 2017/2018, EXPERIENCE OF THE LIGURIAN CENTER AND COMPARISON WITH NATIONAL DATA.

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Lymphedema is a particular pathological disease characterized by a state of tumescence of soft tissues, usually superficial, due to the accumulation by stasis, in the interstitial space, of high protein liquid, the lymph, which has its origin in the reduction the transport capacity of the lymphatic system, determined by the obstruction or abnormal development of the lymphatic vessels and / or lymph nodes of the affected body district.

Lymphedemas that do not recognize a clearly identifiable cause (lymphedemas also called idiopathic) are defined as "primary", although it is often possible to identify one or more "triggering" etiological factors. Generally, this category also includes "congenital lymphedema proper", so defined because it is already manifest at birth. Sometimes congenital lymphedemas recognize an heir-family base (M. di Nonne-Milroy), which can be found, but not constantly, in the evidence of a chromosomal anomaly.

Exceptionally, a congenital lymphedema can follow an amniotic bridle or be associated with other congenital pathologies (for example, when localized to a lower limb, a dislocation or subluxation of the hip).

From a physiopathological, pathogenetic and anatomo-pathological point of view, primary lymphedemas can be reflected in pictures of hypoplasia, hyperplasia, lymphatic-lymph node dysplasia and impaired lymphogenesis.

The developmental anomalies of the lymphatic circulation can specifically affect the lymph nodes, the lymphatic vessels or both such orders of structures. But we must keep in mind, however, on the guidance of clinical experience, that, in most cases of hypoplasia, it is possible to demonstrate an almost constant involvement of the lymphatic ganglia, such as to justify the completely overlapping obstructive picture, on the pathophysiological and diagnostic with lymphoscintigraphy, to that of the classic lymphedema secondary to lymphadenectomy and / or radiation therapy.

Hypo- and lymphatic-lymph node dysplasia can be numerical and structural, in relation, that is, to the number of lymph nodes and / or lymphatic collectors of the district affected by the disease, or to the structure of the vessels (parietal and / or valvular) and ganglia, representing about 90% of cases of primary lymphedema.

Hyperplasia, however, which mainly affects the number of lymphatic collectors, increased abnormally in the district concerned, is also generally associated with lymphatic-lymph node structural dysplasia and corresponds to about 8-10% of cases of primary lymphedema.

The hypo-dysplastic base of many lymphedemas c.d. Primaries can also be reflected in the reduced ability to form and activate an adequate collateral circulation, in the circumstances in which this replacement is necessary (for example, following trauma, infections, surgical interventions, etc.).

It is equally important to take into consideration the alterations of lymphogenesis, as well as those of lymphodynamics.

Lymph formation may, in fact, be abnormally increased or reduced. A state of "hyperlinphogenesis" can result, for example, from the existence of pictures of district arteriovenous hyperstomy or arteriovenous fistulas (such as, for example, in Klippel-Trénaunay disease) or related angiodysplasias (RC Mayall, 1972) .

The condition, however, of reduced or lacked lymphogenesis, due to agenesis, hypoplasia or altered permeability of the wall of the so-called "initial lymphatics" (or lymphatic capillaries), represents a very rare, if not exceptional occurrence.

Finally, among the lymphodynamic alterations, apart from those due to insufficient lymph drainage along the anatomically pre-established pathways, to which reference has already been made, it is essential to also report the reflux of lymph and / or kilo from top to bottom, due to incompetence o insufficiency of the intrinsic anti-gravity means, normally represented by the valves, by the reticulo-myo-elastic structure of the wall of the lymphatic collectors and by the structural architecture of the lymphatic ganglia themselves ("gravitational reflux lymphedemas and chyledema").

With the introduction of the new LEA, lymphedema in its primitive form is included in the RARE disease register with the specific exemption code RGG020.

The regions in compliance with the introduction of the new LEA had to establish which centers belonging to the NHS could certify the patients affected by Lymphedema, this unfortunately in some regions was arbitrarily attributed to certification centers already for other pathology, but it did not always correspond to a real one specialization in the treatment of lymphedema.

In this study, the results communicated to the portal of 16 regions, and of the respective centers for a total of 425 certified cases from 2107 to 2018, will be treated. Liguria alone has certified and taken charge of 81 patients, equal to over 20% of nationally certified patients.

CIRCUMFERENTIAL MEASUREMENTS TO CALCULATE LOWER LIMB VOLUME IN PERSONS WITH LYMPHEDEMA. WHAT SEGMENT LENGTH IS TO BE RECOMMENDED?

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Introduction: Fewer CM than every 4th cm in lower limb volume (LLV) measurements would shorten the measurement time but knowledge about this is currently lacking. The **aim** of this study was to establish the agreement between LLV measurements derived from CM every 4th cm (V4) with every 8th cm (V8) and every 12th cm (V12), and to evaluate the intra-rater test-retest reliability for each of the three measurement methods in persons with lower limb lymphedema (LLL).

Methods & Results: Forty-two persons with unilateral or bilateral LLL were measured twice, two weeks apart using the V4 method. Volume measurements for the V4, V8 and V12 method was derived using CM from the first test occasion. The agreement was evaluated using intra-class correlation coefficient (ICC) and Bland-Altman graph. The intra-rater test-retest reliability was evaluated using ICC and measurement errors (standard error of measurement (SEM%) and smallest real difference (SRD%). The agreement was very high for the V4 and V8 method (ICC 0.999) and for the V4 and V12 method (ICC 0.997). The graphs revealed slightly higher agreement between the V4 and V8 than between the V4 and V12 method visualized by the narrow 95% limits of agreement (LOA) (-129-65 ml and -243-141 ml, respectively). For all three measurement methods, the test-retest reliability was high (ICC 0.992-0.995) and the measurement error low (SEM%: 1.2%-1.5% and SRD%: 3.4%-4.1%).

Conclusions: The faster V8 method for LLV measurements can be recommended in persons with mild to moderate LLL instead of the 'gold standard' V4 method.

PEROMETRIC 3D SCAN IN THE MANAGEMENT OF CHRONIC LYMPHEDEMA

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Topics: Lymphedema management, transition phase, Lymphedema follow-up

Background: The complex decongestive therapy (CDT) is considered the gold standard for the management of lymphedema. CDT includes two phases: intensive phase (IPh) which aims is to reduce the volume of lymphedema and maintenance phase (MPh) to preserve the result. In the intensive phase, is used a complex decongestive therapy, mainly multicomponent high stiffness bandage and manual lymphatic drainage (MLD) associated or not with IPC (intermittent pneumatic compression). In the maintenance phase, we used elastic compression (stockings). The timing of this important transition is often linked to the individual experience of the doctor who can not rely on objective evaluation element. Thus, he risks to place the elastic stocking too early, with a relative reduction in patient compliance or even partial therapeutic failure.

Aim of the work: Define a "percentage range" of limb volumetric reduction, to determine indicatively the time interval necessary to move from the attack phase (bandage use) to the maintenance phase (use of elastic stocking) of the therapy.

Material & Methods: Optical three dimensional (3D) volumetric measurement system (Bodytronic® 600) was used for assessment of lower limb circumference and volume.

During the IPh the Maccio Lymphatic Bandage (LyMB) was used: a highly rigid multilayer bandage. Standard elastic stockings were used during the MPh.

Results: It has been observed that a progressive reduction in the volume of the limb of about 10% compared to the initial volume indicates the achievement of stabilization of the limb or the time beyond which further bandages would not significantly change the situation. This moment is reached about twenty days after the beginning of the therapy and therefore indicates the passage from the bandaging phase to the stocking one.

In addition, the 3D perometer has proven to be a very useful tool for patient monitoring. In fact, in addition to semeiotics and tissue ultrasound we have no other non-invasive means to check the patient's clinic (both lymphoscintigraphy and lymphography and lymphatic MRI still require costs and compensation that are often not manageable on an outpatient basis). With the analysis of over 8000 scans we can say that we have come to have a large surveyed case series that can constitute a reference for the management of the various clinical situations of the patient with lymphedema.

IMMUNOHISTOCHEMICAL OBSERVATION OF THE MICROVASCULATURE OF THE HUMAN MAMMARY TISSUE

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Objective: To elucidate the structure of microvasculature including blood and lymph vessels of the human mammary tissue.

Materials & Methods: The material of the study is surgically resected breast tissues. They were made into FFP-sections and subjected to immunohistochemical reaction using anti-podoplanin, anti-von Willebrand factor, anti-CD34, and anti-CD31 antibodies.

Results: The lymphatic capillaries distributed in the outer peripheral zone of the mammary gland. In the mammary gland the lymphatic capillaries distributed around major and interlobular mammary ducts. In addition, between the major mammary ducts, a rich distribution of lymphatic capillaries in connective tissues was present. There was lymphatic capillary distribution in perilobular region but there was no intralobular distribution.

The blood vessels distributed in the outer peripheral zone of the mammary gland sometimes in conjunction to lymphatic capillaries. In the mammary gland the blood capillaries distributed around major and interlobular mammary ducts. There was blood capillary distribution in perilobular region and in intralobular region around interlobular mammary ducts.

Conclusions: Both blood vessels and lymphatic capillary distributed in periphery of mammary gland, around major and interlobular ducts, and around mammary lobules. But, in the mammary lobules only blood capillaries are present, and no lymphatic capillary distribution was demonstrated there.

REGIONAL LYMPHEDEMA - THE LYMPHATIC DRAINAGE SYSTEM MAY BE DAMAGED LOCALLY IN VARIOUS LOWER LIMB REGIONS BUT NOT NECESSARILY IN THE WHOLE LIMB

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Objectives: Foot and calf skin infections, blunt trauma and fractures are the most common risk factors for long-lasting soft tissue inflammation leading to fibrosis, edema fluid leakage and ulcers, sporadically sepsis. The first hidden symptom of tissue changes is persistent palpation pain and hot spots of calf tissue. These are the early signs of inflammation requiring visualization of the subdermal events and initiate therapy.

Aim: To perform ICG visualization of foci of lower limb inflamed tissues, lymphatics and lymph nodes in patients complaining of foot or calf pains, hot spots and sporadic edema.

Materials & Methods: One hundred randomly selected patients reporting painful calf tissues and sporadically edema were included. Excluded were patients with acute inflammatory symptoms, venous thrombosis, profuse varices, obesity, cardiac insufficiency edema, The ICG visualization of spreading dye in tissues, lymphatics and nodes was done after injection of 0.4ml 5% ICG into toe webs of both feet and one hour walk. The leg image was divided into 15 areas where lymphatic collectors are located and drainage from them was analyzed.

Results: In 90% of obtained images pathological changes were detected. They were: confluent spread of dye in foot dorsum, dilated foot and calf lymphatics, spotty foci in lower calf, dilated thigh lymphatics, confluent spread in thigh and groin, outline of small inguinal nodes or their lack. Most common were changes were detected in the foot dorsum and medial aspect of lower calf. Decreased area of inguinal lymph nodes was observed in the affected limb. Popliteal nodes were depicted in ca. 50% of patients. The leg images were classified as a. dense, b. foggy, c. reticular, d. spotty and e. net web. The area of images was consistently larger than that painful and hot.

Conclusions: The ICG tissue images detect clinically non-diagnosed inflammation foci in the lower limb soft tissues at sites predisposed for lymphedema formation, fibrosis and lymph leakage. A hint for early therapy.

ICF AND DISABILITY.

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According to the World Health Organization (ICIDH 1980), Rehabilitation must be understood as: that process of problem solving and education in the course of which a disabled person is brought to reach the best possible standard of living on a physical, functional, social and emotional level, with the least restriction of his choices, although in the context of the limitation of his impairment and the quality of the available resources. Process that for serious disabilities must also involve the family of the disabled person and those close to him.

Taking charge in rehabilitation is compulsorily carried out through the implementation of an individual rehabilitation project which represents the set of propositions elaborated by the rehabilitation team.

The rehabilitation project is carried out through single interventions that represent therapeutic rehabilitation program.

The Rehabilitation Project generally defines some short, medium and long-term objectives, the expected times, the actions and the conditions necessary to achieve the desired outcomes.

Who identifies the Goals? The International Classification of Functioning, Disability and Health (WHO 2001).

Measuring the person's needs and the assessment of their clinical conditions, integrated with the assessment of available resources and the person's living environment, identifies the many Goals that will lead to the final global Outcome.

The ICF offers us the Bio-Psycho-Social Model of Medicine and Rehabilitation. It is the state of health derives from the interaction between a certain health condition and an unfavorable environment

The ICF is the classification of the state of individual's health which is composed of the assessment of the 4 areas of which the person is formed: Body Structures, Body Functions, Activities Performed, Life Environment.

It is made up of more than 1800 Items describing the person. Their evaluation and qualification describe the person: how he lives, how he relates. This evaluation in pathological conditions describes what the people is unable to do and therefore shows the goals that rehabilitation must achieve in order to reach the final outcome which consists of a functional outcome and a social outcome.

In order to facilitate the development of this path, the WHO has codified Core Sets for some pathologies. One is dedicated to lymphedema.

The core set for lymphedema consists of 3 distinct series of Items: one for the individual suffering from lymphedema in the upper limb, one for lymphedema in the lower limb and one for that in the midline of the body. Each of these takes into consideration the individual as a whole and not just lymphedema impairments. There is also a brief form for each of these works made up of about 20 Items each.

The compilation of a core set for a person shows the limitations and consequently the goals of the Rehabilitation. For each identified goal, the Rehabilitator proposes a specific therapeutic package to take it within the foreseen time frame. From goal to goal, Rehabilitation reaches the final outcome.

The author proposes a method to correctly use the Core sets and thus formulate a correct Rehabilitation Project.

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LYMPHOLOGY - ORIGINS AND FUTURE DEVELOPMENT?

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Lymphology is perceived by the professional public as an unclearly defined medical field dealing with the treatment of lymphoedema. Oedema is not a clearly defined disease but merely a symptom of a disorder of one of the body's systems. In this case, the lymphatic system.

Although the lymphatic system plays an important role in the physiology of many body processes, its insufficiency is not given much attention during both undergraduate and postgraduate studies. There are several reasons. The main ones include the lack of standardized high-resolution examination methods, whether imaging, histological or biochemical. Another and perhaps the most important reason for the lack of interest in the lymphatic system and its function is the unfortunate, though historically justifiable, name for the clinically most obvious manifestation of lymphatic insufficiency: lymphoedema.

If we stick to proper medical terminology in connection with the development of the disease, then, expressed in a rather simplistic way, it originates from either primary or secondary causes. Both of these etiopathogenetic factors lead to the development of lymphatic insufficiency, manifested by terminal pathological conditions characteristic of each particular tissue and organ in the form of fibrosis, lipohypertophy, or recurrent or chronic inflammation. From this perspective, lymphoedema is only one of the clinical manifestations of lymphatic insufficiency in the epifascial space, just like lymphorrhea, lymphocysts, lymph fistulas, recurrent erysipelas, skin ulcer and many others. There are also a number of diseases that have been associated with lymphatic system insufficiency in recent years. The most commonly mentioned are atherosclerosis, asthma or idiopathic intestinal inflammations. Syndromes of numerous diseases have also been associated with lymphatic insufficiency.

The historical and persistent notion of the function of the lymphatic system has revolved around its drainage capability, especially of the epifascial compartment, which results in the clinical picture of swelling (lymphoedema) if disrupted. In fact, this function only provides the environment and it is the mediator that serves more important functions, namely **maintaining metabolic and immune homeostasis of the intercellular environment** of virtually all tissues and organs.

By confusing the medically correct description of the lymphatic system function disorder as the "lymphatic insufficiency" with the now obsolete term "lymphoedema", we are closing lymphology as a medical field into a cage. Until lymphology escapes from this cage, it will not be able to develop as a modern medical discipline.

NOVELTIES IN GENETIC STRATIFICATION OF LYMPHEDEMA

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Primary lymphedema can be present at birth, or develop in childhood or later in life. Thus, it may be a developmental disorder and/or due to a dysfunction of lymphatic vessels that develops with time. Since the discovery in 2009 of the first gene mutated in primary congenital lymphedema or Milroy's disease, the VEGFR3 gene, mutations in numerous genes involved in the initial formation of lymphatic vessels (including valves), in the growth and expansion of the lymphatic system and in associated pathways have been identified in syndromic and non-syndromic forms of PLE. Thus, the current hypothesis is that the majority of cases of PLE has a genetic origin, although a causative mutation can be identified in only about one-third of affected individuals. Overall, three patterns of inheritance are observed, including autosomal dominant (including de novo mutations), autosomal recessive and X-linked. Moreover, penetrance is often less than 100%, making identification of mutations more difficult. Most mutations cause loss-of-function, although some, such as in the genes associated with Noonan syndrome, gain-of-function. Various functional pathways seem to be involved. Based on current knowledge, an algorithm dividing lymphatic anomalies into those with underlying hyperplasia or hypoplasia, and associating them with known genetic mutations, gives clues as to possible medicinal drug intervention on some of them. Thus, the era of theragnostics has also entered the field of syndromic primary lymphedema.

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Friday, June 17th, 2022

SESSION 7: POSTER

Chairs: Boccardo F., Cestari M.

THE EFFICACY OF REHABILITATIVE AND WOUND HEALING APPROACHES IN A MORBID OBESE PATIENT WITH LYMPHEDEMA

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Introduction: The management of advanced lymphedema with wounds is difficult especially in obese patients. Herein we report a morbid obese patient with advanced lymphedema and extensive wounds who was successively treated with combined rehabilitation and wound healing programmes.

Method: A 36-year-old male patient (BMI:54) was admitted to our hospital with swelling in right lower extremity, difficulty in performing activities of daily living (ADL) and mobility. His complaints started after a lipoma excision surgery from right inguinal area but increased significantly in the last one year. Also, he had cellulitis six times in the last one year. He did not have any other co-morbid disease. The patient had advanced lymphedema with characteristic skin changes, skin folds, lobular lymphatic accumulations and extensive wounds. The patient was diagnosed with stage 4 lymphedema and hospitalized for lymphedema rehabilitation and wound healing treatment. Replacement therapy was provided for deficiency of vitamin-D and B12. Treatment plan of the patient for extensive wounds was established with wound-healing unit of our hospital. Systemic antibiotherapy regimen was applied for treatment of infective wounds. Also local antifungal and antibacterial creams were applied. Micronized flavonoid fraction tablets were continued during treatment. A special slimming diet was started. Complete decongestive treatment (CDT) consisting of skin-care, manual lymphatic drainage, multilayer bandaging and therapeutic exercises were applied for 6 weeks and the treatment of patient is still going-on.

Results: The volume of right lower extremity was decreased by 26.5%, wounds were partially healed and he lost 14 kg at the end of 6 weeks. His mobility increased and became more independent in ADL. His quality of life improved in addition to reduction in depressive mood.

Conclusion: Our case report indicates that even a morbid obese patient with extensive wounds can be successively managed and treated with intensive multidisciplinary rehabiliation program and application of appropriate wound healing techniques. In conclusion we would like to emphasize the importance of combined multi-modal therapies in morbid obese patients with advanced lymphedema and wounds.

CONGENITAL LYMPHEDEMA IN A PATIENT WITH PROTEIN C DEFICIENCY AND HYPOXIC BRAIN INJURY

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Background: The diagnosis and management of lymphedema is difficult in patients with thrombophilia. Herein we report a patient with congenital lymphedema, protein C deficiency-a thrombophilic disease- and hypoxic brain injury.

Case Report: A 18-year-old male patient was admitted to our hospital with hypoxic brain injury and protein C deficiency. He had hypoxic brain injury after contrast agent infusion when he was 8 months old. He was diagnosed with protein C deficiency and he was under hematology deparment follow-up since that time. He was wheel-chair dependent. He did not have any heart or renal disease. He had swelling in lower extremities in the last few years in both thigh regions. On physical examination, he had stage 3 lymphedema in both thighs. Doppler ultrasonograpy revealed normal venous and arterial flow in both lower extremities. Also ultrasonographic findings were concordant with lymphedema. The patient received complex decongestive therapy (CDT) comprising; skin-care education, manual lymphatic drainage, multilayer bandaging and exercises for 2 weeks as a total of 10 sessions.

Results: The volume of both lower extremities decreased by 10% at the end of CDT treatment. Manual lymphatic drainage massage and exercises were continued by care-giver of the patient.

Conclusion: Hematologic diseases like protein C deficiency may lead to thrombosis related extremity swelling. Doppler ultrasonography is an important diagnostic technique to rule out thrombosis before diagnosing lymphedema. CDT is an effective treatment method for lower extremity lymphedema and should be initiated promptly after diagnosis of lymphedema.

LYMPHATIC CORD FOLLOWING AXILLARY NODE DISSECTION

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Introduction: The axillary cord (AC) phenomenon occurs in the axilla and on the frontal side of the upper arm and sometimes along the forearm to the thumb. The cord is painful, in particular by movement, and can therefore be very disturbing to the patient. Though the phenomenon has been examined and discussed for decades, no evidence for the origin has been found until now. The aim of this study was to perform a histopathologic analysis of cords taken 1996-98 in the Surgical Clinic, Skane University Hospital, Lund.

Material & Method: In seven patients with AC, biopsies of the cord were taken 4-5 weeks after axillary node surgery due to breast cancer, and analyzed for endothelian cells of lymphatic vessels using the D2-40 method.

Results: In one biopsy, there is a dilated vessel with a thickened wall which is confirmed by D2-40 immunostaining to represent a lymphatic vessel. The lumen is occluded by organised thrombus within which new vessels are being formed indicating recanalisation. In two other biopsies, similar lymphatic vessels with thickened walls are also present although the lumen of the vessels are not visualised in the planes of section. The other four biopsies do not show specific features.

Conclusion: Although only one case this is the first pathological evidence of thrombosis within a confirmed lymphatic vessel from a case of cording. We propose that AC represents lymphatic vessel thrombosis. Recanalisation of the thrombus eventually restores lynmphatic flow consistent with the transient nature of the condition.

CELLULITIS (DERMATO-LYMPHANGIO-ADEMITIS, DLA) IN LIMB LYMPHEDEMA – ANTIBIOTIC ACUTE AND CHRONIC THERAPY, PROPHYLAXIS

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Objectives: Dermato-lymphangio-adenitis (DLA) affects about 50% of patients with obstructive lymphedema of lower and upper limbs. Each recurrence is followed by progression of edema and irreversible increase in limb size. The question arises how to effectively treat acute cases and those with recurrency, as well as, how to prevent its development in patients after surgical oncological treatment.

Aim: To standardize antibacterial policy toward acute and chronic bacterial inflammation in the lymphedematous limbs and prevention of inflammation development after oncological procedures.

Methods: Study was carried out on over 1000 patients with obstructive lymphedema of lower and upper limbs reporting in group I the first acute DLA episode, in group II DLA prior to visit us and reporting more than 3 recurrent DLA attacks and in group III the post-mastectomy and hysterectomy cases with no DLA. In group I amoxicillin, metronidazole and cefuroxime p.o. for 3 days, continuing amoxiclav for 21 days, and simultaneously applying one shot of 1200000u of benzathine penicillin every 14 days to be continued for 12 months. In group II 1200000u of benzathine penicillin was given every 14 days for minimum 12 months, usually carried on for years. In group III all patients received 1200000u of benzathine penicillin every 14 days for a minimum of 5 years. Frequency of DLA recurrency was observed for 5 years, although in a large number of cases it was extended to over 10 years.

Results: Group I. The given therapy was effective in all patients followed for 1 month. Most patients joined group II as they were further treated with benzathine penicillin. Group II. Serum penicillin concentration was maintained above 20 pg/ml. No DLA recurrency occurred in 90% of patients over a mean 60 months of penicillin administration. In the remaining 10% decrease of attacks/person was by 55%. In group III. No statistically significant incidence of DLA was observed.

Conclusions: We presume that permanent presence of slowly absorbed penicillin keeps the tissue dwelling bacteria in a non-proliferating state. We consider years-long administration of long-term penicillin an obligatory condition in all cases of obstructive lymphedema after an experienced DLA episode

THE LOWER LEG CONFINED INFLAMMATION IS GRADUALLY FOLLOWED BY DAMAGE TO LYMPHATICS AND DEVELOPMENT OF WHOLE LIMB LYMPHEDEMA

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Objectives: Circumscribed foci of inflammation of the calf are seen more frequently especially in obese patients. This is why we see more isolated lymphedema regions limited to foot or part of calf only. On the ICG imaging confluent areas of dye at site of inflammation with preserved flow in the dilated collectors can be seen. Early diagnosis and appropriate antibiotic treatment of foot or calf infection may limit spread of microbes and the region of inflammation. Early imaging of inflammatory areas by ICG lymphography may help in initiation of a fast therapy.

Aim: To perform ICG lymphography in patients complaining of circumscribed inflammation and edema of any part of the lower limb so far not diagnosed as lymphedema, and apply1200000u of benzathine penicillin every 14 days for minimum 12 months, in a number of cases carried on for years.

Material & Methods: One hundred of patients showing up in OPD with circumscribed inflammatory regions of foot and/or calf, with no clinically evident edema of the limb, underwent ICG lymphography.

Results: 1. Foot and lower part of calf erythema: confluent spread of ICG in the dorsum and plantar area, outline of collecting lymphatics in the calf and thigh and enlarged inguinal nodes. 2. Calf erythema. Outline of dilated foot lymphatics, confluent spot embracing lower calf and slight outline of thigh lymphatics with enlarged inguinal nodes. Therapy. Evident decrease of erythema was observed in all cases. Post-treatment ICG lymphography. In patient agreeing to undergo the follow up lymphography images didn't differ from those obtained before treatment, however, there was no increase in the size of erythema or obstruction of collectors

Conclusions: Early ICG lymphography depicts areas of lymphatic involvement by the inflammatory process. Immediately employed therapy may prevent spread of damage to collecting trunks and nodes

LYMPHATIC COMPONENT OF LEG VENOUS ULCERS – LYMPHOGRAPHY EVALUATION OF THE SIZE OF PERI-VENOUS INFLAMMATION

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Objectives: The lymphatic system participates in leg tissue inflammation, wounds and ulcers by transport away of microbial and tissue antigens to the lymph nodes, where from phagocytic and cytotoxic cells are dispatched and migrate to the ulcer. In the non-healing venous ulcers lymphatics are persistently involved. This process has not been studied until recently as there were no sensitive imaging methods of lymphatics in the inflamed tissues.

Aim: To evaluate the ICG images in legs with venous ulcers and specifically size of peri-ulcer inflamed regions, lymphatic drainage from ulcer bottom and enlargement of the draining inguinal lymph nodes.

Materials & Methods: Twenty patients with the non-healing calf venous ulcer (CEAP 6) of over 1 year duration were included. Indocyanine green (ICG) 5% 0.1 ml was injected into a) toe-web and additionally after one hour below and lateral to ulcer. In some cases ICG was injected into ulcer bed. Images obtained after 2 and 24 hrs were evaluated. As controls served the contralateral normal limbs.

Results: Two hours images. Dilated foot lymphatics reaching ulcer level. Extravasation of ICG around ulcer, no dye in the ulcer bed. The area of ICG foggy picture often encompassed the calf. Twenty-four hour images. ICG spread around ulcer and to the inguinal lymph nodes. Outline of enlarged inguinal nodes.

Conclusions: The ICG lymphography showed peri-ulcer area of increased plasma capillary filtration and immune cell extravasation and inadequate lymphatic collectors capacity to evacuate the excess volume of tissue edema fluid. Moreover, it depicted enlarged lymph nodes. This is for the first time, using lymphography, that venous ulcer is not only circumscribed skin necrosis, but also a large area of tissue and nodes inflammation.

LONG-TERM FOLLOW-UP OF SILICONE TUBE IMPLANTATION FOR BYPASSING THE SITE OF LYMPH FLOW OBSTRUCTION - 2020

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Objectives: We proposed in 2010 to drain edema fluid accumulations in advanced stages of lymphedema with obliterated collecting trunks by creating artificial pathways to non-obstructed regions by implantation of “silicone lymphatics”. The 10 year follow up in around 100 patients provide information of the therapy and side effects.

Aim: To present the 10 years follow up results of therapy of lymphedema by subcutaneous implantation of silicone tubings in lower and upper limbs and fate of tubing as well as complications.

Methods: In over 300 patients in our centers with obstructive limb lymphedema after pelvic or axillary lymphadenectomy and irradiation in uterine or breast cancer or following soft tissue inflammation silicone tubes were implanted subcutaneously. Compression and prophylactic penicillin were applied in the same fashion before and after implantation.

Results: a) immediate decrease of limb circumference within days after implantation b) in lower limbs in 5 year follow up a decrease in mid-calf circumference by a mean -9% (P<.05) with range of -3% to -30 % corresponding to 90-900ml volume and in mid-thigh a mean –2.0% (P<.05) with range of 10% to + 3% equal to 0-900ml. In upper limb decrease in mid-forearm was -8.5% (P<.01) with a range of -3.0% to -25.0 % and in mid-arm a mean –10% (P<.05) with a range of -7% to - 25%, corresponding to 180-700ml volume. These data did not different from those after 10 years. The new observations included a) “dryness” of tissues with no more fluid squeezed out on plethysmography, b) all tubes patent, c) encapsulation of tubing, d) no major inflammatory reactions to tubing in 95% of lower limbs and no such episodes in upper limbs, e) in 5% of cases inflammation appeared after leg trauma, foot abrasion, or barefoot walking. It required intra-tubing antibiotics or removal. Re-implantation of tubing into other tissue channel was done in 16 patients.

Conclusions: Treatment of advanced lymphedema by silicone tubing implantation has become a routine effective procedure. Long term observations point to inflammation after leg abrasion and encapsulation of tubing requiring re-implantation.

LYMPHEDEMA MAY BE CAUSED NOT ONLY BY LYMPHATIC OBSTRUCTION BUT ALSO BY LOSS OF LYMPHATIC CONTRACTILITY

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Objectives: Early stage of lymphatic insufficiency in human limbs can now be observed on the near infrared ICG lymphography. It differs from “lazy” contractions to total stop and passive lymphatic filling by external massage. Once visualized on ICG lymphography, lymphatics are seen contracting autonomically depending on filling up of lymphangions (Olszewski PTL 1968, AmJ Physiol 1980). Their contraction (pulse) capacity as frequency, pressure amplitude and stroke volume can be measured. After inflammation, trauma and lymph hypertension due to proximal collector obstruction tissue fluid is not entering lymphatics, forms a spot, collectors are filled with dye only after compression.

Aim: To evaluate foot and lower calf collector contractions and lymph flow capacity in initial stages of lymphedema and evaluate semi-quantitatively on fluorescence level curves.

Methods: Fifty patients with lower limb lymphedema stage I underwent ICG lymphography procedure in a horizontal position with 3 minutes foot flexion (first recording) followed by one hour walk (second recording). Time of dye entry to foot lymphatics, appearance of foot lymphatic outline, lymphangion contraction rate at rest and after 3 min foot flexion/extension, appearance of calf lymphatics and diffusion of dye across the non-contracting collectors were recorded. The obtained curve pulse waves and area of fluorescence changes in time were analyzed (ICcalc, Pulsion Medical Systems, Munich).

Results: Low pulse frequency (below 6/min) and amplitude was usually observed in one or two lymphatics with other dilated non-contracting collectors as well as distal spread of ICG, compared with healthy limb (fluorescence 30% vs 15%). A variety of individual images was observed but common to all was slowing down of dye flow.

Conclusions: Evaluation of contractility of foot and calf lymphatics is now possible on ICG lymphography. This enables early diagnosis of lymphatic insufficiency and initiate prophylactic procedures

PECTORALIS MAYOR RUPTURE IN A PATIENT WITH BREAST CANCER. SECONDARY LYMPHEDEMA. A CASE REPORT

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Introduction: Lymphedema after breast cancer treatment may present immediately or years after treatment, although the majority of cases occur the first two years. Sometimes it appears due to a trigger.

Materials & Methods: We present the case of a 36-year-old woman who underwent surgery on the left breast in 2008, performing radical left mastectomy, lymphadenectomy (2/17), adjuvant treatment with chemotherapy, radiotherapy and hormonotherapy. In 2012 reconstruction is performed with an expansive prosthesis and in 2013 a definitive prosthesis was implanted. She went to the clinic in December 2019 for lymphedema. She had gone to the emergency department in June for sudden chest pain and subsequent edema in left upper limb and upper thorax. She was diagnosed with osteochondritis

Physical examination showed breast prosthesis of normal consistency, left arm pitting edema with a limb difference of 43.5 cm in the bilateral multiple circumference measurements of the arms taken in seven points

She started treatment with CPT after imaging tests requested to rule out tumor recurrence

Breast ultrasound: No signs of recurrence, prosthesis without complications.

Breast MRI: Left mastectomy and reconstruction with prostheses without complications and without signs that suggest recurrence. Pectoral muscle alteration

Muscular ultrasound: Findings compatible with rupture of the left major pectoralis muscle, with retraction of its belly over the parasternal region.

Request assessment by Plastic Surgery: value DIEP flap reconstruction and pectoralis major muscle repair. The patient performed CPT with little improvement.

Conclusions: In the evaluation of patients with a history of breast cancer, we must take into account adjuvant treatments that can influence the development of pathologies such as the one described, whose presentation is unusual.

THE MANAGEMENT OF GENITAL LYMPHOEDEMA

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Genital lymphoedema is a most embarrassing medical problem for the patients so they usually come at a very late stage for treatment at that stage it is very difficult for the surgeon or physician to manage this problem we has developed a new technique of multimodal treatment for this patients both in the male and female patients which includes a new surgical technique for both male and female patients and we have attained a very good post treatment out come with a fallow up of 20 years. Which will be explained with clinical and per operative and post operative pictures in our presentations (poster on video).

BREAST CANCER-RELATED LYMPHEDEMA: RISK FACTORS AND PREDICTION MODEL

MARTÍNEZ-JAIMEZ P.[1,2], ARMORA VERDÚ M.[3], CARLOS G. FORERO C.[4], SAMANTHA ÁLVAREZ SALAZAR S.[4], FUSTER LINARES P.[2], MONFORTE-ROYO C.[2], JAUME MASIA J.[1,3,5]

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Aims: To identify the risk factors for lymphedema following axillary lymph node dissection in a European sample and to propose a lymphedema prediction model for this population.

Design: Predictive retrospective cohort study comparing women who developed lymphedema within two years of undergoing axillary lymph node dissection with those who did not developed lymphedema.

Methods: We reviewed the clinical records of 504 women who, between January 2008 and May 2018, underwent surgery for breast cancer that involved axillary lymph node dissection. Logistic regression was used to identify significant risk factors for lymphedema. The prediction accuracy of the model was assessed by calculating the area under the receiver operating characteristic curve.

Results: Of the 504 women whose records were analysed, 156 developed lymphedema. Significant predictors identified in the regression model were level of lymph node dissection, lymph node status, post-operative complications, body mass index and number of lymph nodes extracted. The prediction model showed good sensitivity (80%) in the study population.

Conclusions: The factor contributing most to the risk of lymphedema was the level of lymph node dissection, and the only patient-related factor in the prediction model was body mass index. The model offers good predictive capacity in this population and it is a simple tool that breast care units could use to assess the risk of lymphedema following axillary lymph node dissection. Nurses with specialist knowledge of lymphedema have a key role to play in ensuring that women receive holistic and individualized care.

ADJUSTABLE COMPRESSION WRAPS SYSTEMS IN SEVERE LIPOLYMPHEDEMA

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Introduction: Lipedema is a chronic adipose tissue disorder with symmetrical fatty swelling accompanied by pain and bruising due to minor trauma. Patients with lipedema may develop secondary lymphedema (lipolymphedema), which may worsen if chronic venous insufficiency is also present.

Assessment to determine which type of compression therapy should include the severity and extent of the lipolymphedema, the presence of skin folds or fat lobes and pain.

Material & Methods: We present the case of a 54-year-old woman with increased volume in lower limbs since puberty. She had a progressive worsening in the last five years below the knee and feet swelling. The patient had limitations in activities of daily living, including walking and stair-climbing.

Initial physical examination showed a BMI of 56.01, limitation in knee joint mobility with a range of movement (ROM) 0º-80º, abnormal fat distribution in the buttocks, thighs with deep folds. She had pitting edema with foot involvement in the region below the knee. She presented skin lesions with signs of bacterial infection in the heels and fingers area. To assess the swelling, we made seven circumferential measures; the sum of them was 435 cm on the left and 404.6 on the right.

She presented a remarkable improvement after performing treatment consisting of antibiotic therapy, bodyweight control and complex decongestive therapy (CDT) with a combination of manual lymph drainage, compression therapy with adjustable compression wraps, exercise and skincare.

After hospital discharge, she presented a BMI of 46.48, knees ROM increased 30º, the sum limb circumference decreased 73,5 cm on the right leg, and 82,6 cm on the left one.

On the subsequent medical consult, we renewal the compression devices using an adjustable compression wrap for the thighs and knee combined with hosiery for the lower leg. After 1.5 years, we changed adjustable compresión wrap for an integrated system of capri pants, below-knee stockings and adjustable compression leg wrap. In the last rmedical review, two years after discharge, the swelling goes on decreases, so finally, the sum limb circumference decreased 101.6 cm on the right limb and 107,8 cm on the left one.

Conclusions: Combining dermal complications treatment, body weight control, and complex decongestive therapy associating compression wraps is very effective for severe lipo lymphedema.

For patients with severe edema with significant shape distortion, adjustable compression wraps may be more accessible and less painful to apply than other types of compression systems. They have the advantage of allowing the patient to adjust the degree of compression and are less likely to cut into soft tissues.

MORPHOLOGICAL CHARACTERIZATION OF ADIPOSE TISSUE FROM PATIENTS AFFECTED BY LIPEDEMA

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Lipedema is a pathology still unknown that mainly affects women of young age, in particular in the third decade of their life. Lipedema is characterized by abnormal bilateral and symmetrical increased subcutaneous adipose tissue present in the limbs, with no involvement of the trunk, the hands and feet resulting a "bracelet" effect at the ankle. While adipose tissue in obese patients does not cause specific symptoms, in patients with lipedema the abnormal increase of adipose tissue causes symptoms such as pain, tenderness, increased vascular fragility, arthritis and easy bruising.
Adipose tissue of patients suffering for lipedema is altered in the specific sites of subcutaneous affected fat, however a specific morphological characterization has not been reported.
We sought to compare adipose tissue of normal weight patients affected by lipedema by comparing the morphology of affected and healthy areas of subcutaneous fat in the same patients.
Two biopsies from each patient were collected, a biopsy from an area affected by lipedema (subcutaneous adipose tissue of the limbs) and a biopsy from relatively unaffected subcutaneous adipose tissue from the sub-scapularis region of the same patient: the second biopsy was used as a control.
We performed: structural analysis by light microscopy observation of Haematoxylin & Eosin or immunohistochemical (PLIN 1) stained sections, morphometric analysis to measure the adipocyte size and ultrastructural analysis by Electron Transmission Microscopy (TEM) observations.
Our results showed, as common feature of affected areas, a peculiar aspect of capillary vessels. They showed hypercellularity either due to endothelial as well as perivascular cells.
The vessels' wall resulted thickened and irregular. When observed by TEM these vessels showed: 1-thickened and reticulated basal membrane; 2-irregular shape with apparent hyperplasia of endothelial cells; 3-presence of several pericytes embedded in the basal membrane; 4-evident signs of calcium deposits.
Adipocytes showed: variable amount of cytoplasmic calcium deposits and frequent signs of degenerative alterations in line with IHC data showing the reduction of immunoreactivity for PLIN 1 in a small % of adipocytes. Abnormal deposition of collagen fibers among adipose cells was constantly observed.
Morphometry data showed an increased size of adipocytes from affected area compared to non-affected area in some patients.
Our data showed that the adipose tissue from normal weight women has substantial morphological alterations. The study needs to be extended and deepened to improve understanding of the pathogenesis of lipedema and provide new therapeutic targets.

DESIGN OF A MULTIDISCIPLINARY PREVENTIVE INTERVENTION FOR BREAST CANCER-RELATED LYMPHEDEMA: AN INTERNATIONAL CONSENSUS

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Objective: To design a consensus regarding the multidisciplinary prevention of breast cancer-related lymphedema (BCRL), taking into account the expert opinion of professional groups from across the world involved in the identification and treatment of breast cancers.

Methods: International consensus study involving a modified nominal group and Delphi process. A total of 50 preventive strategies representing those used by a range of health disciplines involved in breast cancer care were identified by the nominal group. These strategies were categorized into four sub-groups (General recommendations, Therapeutic approach, Rehabilitation and oncological physiotherapy, and Dietary recommendations) and will present in survey format to a multidisciplinary panel of experts in a two-round Delphi process. Eleven specialist areas and fifteen countries will represent on the panel.

Results: A total of 14 experts were involved in the nominal group (NG) process. Participants identified the key strategies for preventing BCRL from the perspective of their specialist area, and this was followed by discussion among the group as a whole regarding points of agreement and disagreement. The steering group then grouped the responses obtained into four blocks, based on their content: 1) General recommendations, including information that patients need for adequate self-care; 2) Therapeutic approach, grouped according to specialist area: oncology, radiology, nuclear medicine, surgery, and radiotherapy; 3) Rehabilitation/oncological physiotherapy; and 4) Dietary recommendations. The preliminary list of 50 strategies for preventing BCRL was then sent to all 14 participants in the NG for appraisal prior to its use in the Delphi process. Ten replied and confirmed the suitability of the survey format and its contents.

Conclusion: We report a design and pre recommendations regarding an international consensus for the multidisciplinary prevention of BCRL, setting out recommendations and clinical guidelines aimed at systematizing care.

IMPACT OF LOCKDOWN ON PPL IN ITALY: FOCUS ON THE EFFICACY OF EDUCATIONAL LYMPHOEDEMA CHILDREN'S CAMPS IN DAILY SELF MANAGEMENT

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Introduction: During Spring 2020 strong restrictions on mobility were imposed in Italy due to Covid-19 pandemic. Access to physical treatments for several chronic diseases, such as lymphoedema, was either discointinued or restricted to acute cases. However, in order to limit the worsening of lymphoedema, iterative physical therapies were required toghether with techniques of self-management for patients and their relatives. Lymphedema children's camps were aimed at promoting joining of families and reducing psychosocial impact of disease. Camps also favored involvement in self care of an increasing number of families.

Aims of study: The aim of this study was to explore the parental experiences of caring children and adolescents with lymphoedema during 2020 lockdown in Italy and to measure the efficacy of education therapy (acquired during children's camps) at the time of pandemic.

Methods: Thirty-nine families of kids and adolescents anonymously participated in an online survey. The mean patient age was 9 years (range 1-19). Twenty-three families (57%) were previously involved in our educational camps (and represented the study group). Data were analyzed using SPSS Statistics.

Results: The evidence of a greater effectiveness of the self care techniques during the lockdown have been proved by the observation that lymphoedema was found to be clinically stable or even improved in the study group compared to controls (Spearman Rho coefficient 0.61; p=0.02). Self-care learning was also associated with a wider variety of employed techniques. A higher propensity in the application of lymphatic drainage vs compression was noted in the study group (Rho 0.53 p.< 0.01). Self caring attitude was also found to be associated with both quality of attendance to educational camps (Rho 0.63, p.=0.02) and number of attendances (Rho 0.57, p.<0.01).

Conclusion: Lymphoedema children's camps had a significant impact on systematic application of self care techniques for lymphoedema management at the time of pandemic.

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Saturday, June 18th, 2022

SESSION 8: CLINICS

Chairs: Borman P., Campisi C. C., Pissas A.

FAT DEPOSITION AS A RESULT OF INFLAMMATORY RESPONSE IN EXPERIMENTAL LYMPHEDEMA

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Increased limb volume in patients with chronic lymphedema is due to tissue fluid retention, fibrosis and fat deposition. The Inflammatory component of lymph stasis has long been recognized whereas scanty attention has been given to fat deposition and its exact mechanism remains unclear.

Recently, a modified model of experimental lymphedema in mice tails provided a suitable model for the study of many physiopathological aspects of lymph stasis.

We evaluated the time course of lymphedema formation, three and six weeks after surgical interruption of both superficial and deep lymphatic pathways in this mouse-tail model and observed a marked increase in the inflammatory mediators(TNF-α, IL-6, MCP-1) and the subsequent infiltration of immune cells (identified by the increased F4-80). Also, adipocytes accumulated the lymphedema area of the tails, initially with an early increase in the expression of C/EBP, which is a fundamental constituent of the adipocyte differentiation process and enhanced expression of genes related to adipocyte maturity was observed later.

To ascertain the causal relationship between inflammation and adipogenesis, we manipulated the inflammatory response in order to determine this relationship by using dexamethasone and mice deficient in NOS2. In both treatment groups there was a marked decrease in inflammatory infiltrates, followed by a significant reduction in mature adipocytes.

The modified mouse tail model of lymphedema showed that inflammation precedes adipogenesis and that MCP-1 and nitric oxide may be potential targets for future therapies for lymphedema.

THE COMPARATIVE FREQUENCY OF BREAST CANCER-RELATED LYMPHEDEMA DETERMINED BY PEROMETER AND CIRCUMFERENTIAL MEASUREMENTS: RELATIONSHIP WITH FUNCTIONAL STATUS AND QUALITY OF LIFE (PRELIMINARY REPORT)

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Objective: The aim of this study was to comparatively determine the frequency of subclinical lymphedema by using prospective monitoring with perometer and circumferential measurements in a group of patients who underwent breast cancer surgery. We also aimed to evaluate the relationship between volume-changes and functional-status and quality of life (QoL) in patients with breast cancer-related subclinical lymphedema.

Material & Methods: A-hundred patients who had unilateral breast-cancer surgery were assessed with circumferential and perometer respectively for volumes at baseline, 3rd-month, 6-month, 9-month and 12-month by the same physiotherapist. Every patient was informed about lymphedema symptoms and prevention. The demographic and clinical properties were recorded. Functional status and QoL was evaluated by Q-DASH and LYM-QOL-Arm questionnaires. The threshold for subclinical lymphedema was determined as >5% excess-volume difference compared to unaffected arm.

Results: 32 female-patients with a mean-age of 50.50+10.50 years completed the 12-month follow-up. Most the patients were over-weight and had axillary dissection in addition to breast surgery. The frequency of subclinical lymphedema at the end of 12-months, was determined by 41% and 22% with circumferential-measures and perometer, respectively. Patients with subclinical lymphedema determined by perometer, were found to have higher BMI and higher number of dissected axillary lymph nodes. QoL and functional scores did not differ between patients with and without subclinical lymphedema.

Conclusion: After the first 12-months after surgery; the frequency of subclinical lymphedema assessed by circumferential measurements was higher than the frequency assessed by perometer. Periodic monitoring of women with breast cancer for presence of subclinical lymphedema, preferably by the same person together with patient education is suggested in order to allow early detection and timely intervention for BCRL.

EVALUATION OF SLEEP QUALITY, DEPRESSION AND QUALITY OF LIFE IN PATIENTS WITH BREAST CANCER RELATED LYMPHEDEMA

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Introduction: Breast cancer related lymphedema (BCRL) is a serious health condition negatively affecting psychological state, sleep quality and quality of life of patients. These complications of BCRL can be overlooked in routine clinical practice. The aim of this study is to evaluate sleep quality, depression and quality of life in patients with different stages of BCRL.

Methods: This cross-sectional study included 111 consequtive women patients with BCRL attending to lymphedema unit of Physical Medicine and Rehabilitation Hospital in Ankara City Hospital. Demographical and clinical data of patients were noted. Sleep quality was assessed with Pitsburgh Sleep Quality Index, day-time sleepiness was assessed with Epworth Sleepiness Scale, depression was assessed with Beck Depression Questionnaire, quality of life was asssessed with LYMQOL-arm.

Results: Median age of patients were 57(49-63) (25%-75% interquartile range). Nearly 58.8% of the patients had sleep disturbances and 29.7% of the patients had depression. Presence of both sleep disturbance and depression were associated with increased age. Patients with sleep disturbance were found to be more depressive and vice versa. Sleep disturbance, depression and day-time sleepiness and higher body mass index were all associated with lower quality of life scores.

Conclusion: Presence of sleep disturbances and depression should be evaluated in every patient with BCRL with special attention to elderly. Rehabilitative management of BCRL should include psychiatric evaluation and interventions. Future studies should intend to assess the risk factors that affect psychological state, sleep quality and quality of life in patients with BCRL.

"SCROTAL LYMPHOEDEMA AND PREPUTIAL ELAPHANTIASIS: CASE REPORT"

SCIUSCIO M.

Servizio territoriale del Linfedema Primario e Secondario-Presidio di Rete Regionale Malalttie Rare Linfedema Primario Cronico. U.O.C. Sovradistrettuale della Riabilitazione ASL Lecce

Introduction: Lymphedema mostly involves the upper and lower limbs but, exceptionally in industrialized countries, it can appear on the external genitalia, configuring a severe condition of preputial and testicular elephantiasis. This clinical picture generates all kinds of disabilities, motor, voiding and psychological with unfavorable repercussions on the patient's quality of life.

Materials & Methods: A 67-year-old patient, he comes for an examination due to the appearance of edema in the left testicle and preputial with a worsening trend which occurred a month earlier. In anamnesis, in 2015, high grade papillary urothelial carcinoma of G2-G3 bladder treated with intravesical immunotherapy with BCG and Monoclonal Gammapathy of uncertain significance (MGUS) in follow-up; subsequent endoscopic urological checks negative for disease recurrence. Testicular and preputial edema had already appeared a year earlier, subjected to a lymphological examination which, without ascertaining its etiopathogenesis, directed him to the treatment of L-V anastomosis (1) in the belief that it was a late-onset primitive lymphedema. The lymphedema then disappeared, within a month, completely spontaneously, leaving only a left hydrocele subjected to hydrocelectomy. Having come to our observation, it is therefore investigated with blood tests, ultrasound examination and PET-TC, lymphoscintigraphy, lymph node cytological examination (3) in order to understand its cause. Meanwhile he is directed to the decongestant treatment for which he performs sessions of multilayer bandage with short-stretch bandages to the external genitalia and manual lymphatic drainage that allow the patient to contain the edema and reduce the psychological and motor discomfort (2) deriving from it. temporarily and with unfortunately reversible results.

Results: Since all the diagnostic tests pointed towards a tumor process in the testicular area with PET-CT evidence of high glucose consumption in the external iliac sinus, inguinal and bilateral femoral lymph node, as well as at the level of the left testis, lymphoscintigraphic evidence of slowed superficial lymphatic drainage at the level of the left lower limb with complete normalization in late images, a lymph node biopsy was necessary also in the light of a non-diagnostic lymph node cytology and an inexplicable and progressive anemia. This is high-grade Non-Hodgkin's Lymphoma with associated haemolytic anemia from underlying lymphoproliferative disease. The patient is currently undergoing chemotherapy with unstable compensation.

Conclusions: Lymphedema is an insidious pathology and often the etiological diagnosis is not simple. The clinical history of each patient directs towards the diagnosis, diagnostic confirmation is however instrumental and, in doubtful cases, a lymph node biopsy is always decisive even in those situations in which we find the operating surgeon disagree, convinced that the excision of the lymph node, may further worsen the clinical picture.

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LYMPHATIC VESSELS SENESCENCE: CURRENT EVIDENCE AND IMPLICATIONS FOR CLINICIANS

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The senescence of body structures leads to anatomical and functional modifications, which may have possible important repercussions on the health of persons.

Lymphatic vessels play important roles, including the homeostasis of body fluids and the innate and adaptive immune response. Potentially, aging of the lymphatic system could have consequences that can become clinically relevant. However, the role of senescence of the lymphatic vessels is poorly investigated. There is a need to carry out extensive research in this area as it is possible that the aging lymphatic system may actually be the main contributor to the various functional changes that are seen in older persons. For instance, the edema of the lower limbs that occurs in aged people may not only be due to cardiac insufficiency or venous incompetence but may also be due, at least partially, to the senescence of lymphatic vessels.

Examined in this talk will be some studies that focused on lymphatic senescence. These studies, performed mainly in mice, have shown several important alterations in the aged lymphatic collectors and surrounding tissues. These include loss of the extracellular matrix, reduction of the collagen surrounding the lymphatic collectors; marked reduction in glycocalyx; alteration of the waterproofing of the vessel wall; increased oxidative stress, reduced contractile proteins, reduced channels for sodium, potassium and calcium; reduction of the frequency and extent of vasal contraction as well as reduced clearance of bacteria. The effect of lymphatic senescence observed in animal experiments can explain some clinical presentations seen in older persons and perhaps also in subjects in early stages of mechanical insufficiency. Therefore, included in this talk will also be some clinical data from patients by the authors of this paper.

This lecture concludes by stating that there is a need for identification of preventive and curative approaches aimed at lymphatic structures and the extracellular matrix and it emphasize the importance that related problems (obesity and chronic venous insufficiency in particular) should be earlier diagnosed and treated.

A QUESTIONNAIRE BASED SURVEY TO ASSESS THE AWARENESS AMONGST HEALTH CARE PROFESSIONALS ABOUT BREAST CANCER RELATED LYMPHEDEMA.

VAGAL M.

Primary objective: To assess the awareness amongst health care professionals (Onco-surgeons /Medical Oncologists/ Radiation Oncologists, Occupational Therapists and Physiotherapists) about breast cancer related lymphedema with the help of a structured questionnaire based survey.

Background and Rationale: Lymphedema (LE) is a chronic progressive debilitating condition which adversely impact survivorship following breast cancer treatment. Awareness about post breast cancer lymphedema amongst the attending oncologists and rehab professionals is very crucial to its early detection, effective outcome and preventing functional morbidity. To understand the awareness about breast cancer related lymphedema amongst health care professionals a prospective, national, questionnaire based survey was carried out.

Methods: A database consisting of contact details of oncologists was populated through breast cancer disease management group (DMG), Tata Memorial Hospital (TMH), Mumbai and All India Occupational Therapists Association (AIOTA) website. Individual mails containing unique link to the questionnaire, was sent to total 800 health professionals working with breast cancer patients in India from the prepared database. Reminder mails were sent after the initial mail to get maximum responses.

Data collection and analysis: The data collected from the online survey was analysed by descriptive statistical methods using IBM SPSS Statistics for Windows, Version 21.0 (Armonk, NY: IBM Corp. Released2013). Nominal categorical variables was descriptively analysed regarding to the observed frequencies. Central tendency and dispersion measures was used to analyse numerical variables. From 800 clinicians total 90 responded in first mail and 35 responded through repeat mails. Survey found that not all respondents attempted all the 20 questions of the questionnaire. From total of 125 respondents five were medical oncologists, six were surgical oncologists, one was radiation oncologists and 87 were occupational therapists and eight were physiotherapists, and the rest were palliative care physician, nursing, gynaecologists, including those who did not specify their professional background. Only 34 professionals had lymphedema management clinic/facility available at their centre. Though lymphedema management related information was found to be poor amongst the professionals (only 17.14%) but 73.75% and 80% professionals reported baseline measurements of both extremities before surgery to be important for early detection and management of lymphedema and 80.56% professionals testified that professionals should be certified to provide lymphedema care. In their routine clinical practice, 78.75% of health professionals considered limb girth measurement as means of lymphedema assessment .Similarly, lack of trained personal (49.23 %) and lack of treatment compliance (36.92 %) was reported as practical difficulties in treating lymphedema in regular clinical practice by the health professionals.

Conclusion: This survey reported not only awareness issues about lymphedema management amongst the health care professionals in India but also lack of adequate infrastructure which is alarming. Awareness issues about lymphedema condition can be addressed by emphasising on application of international best practice guidelines about lymphedema amongst the health professionals in India and in different cancer centres across India. Similarly, national guidelines also may be generated based on international guidelines which may then be circulated to various cancer institutes in India through National Cancer Grid.

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Saturday, June 18th, 2022

SESSION 9: SURGERY

Chairs: Boccardo F., Brorson H., Campisi C., Masia J.

MICROSURGICAL TREATMENT OF BREAST CANCER-RELATED LYMPHEDEMA COMBINED WITH COMPLEX PHYSICAL THERAPY: PRELIMINARY RESULTS OF THE EFFECT ON ARM VOLUME CHANGE

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Objective: To study the effect of microsurgical interventions in combination with complex physical therapy (CPT) on arm volume change in patients with breast cancer-related lymphedema (BCRL).
Hypothesis: The volume difference between the affected and non-affected arm has decreased one year following intervention.
Materials and Methods:

- Design: prospective longitudinal cohort study
- Primary inclusion criteria: BCRL pitting positive (stage I-II), at least 6 months resistant to CPT with an intake at the Universitair Ziekenhuis Brussel between 03/02/2011 and 31/12/2020.
- Intervention: Vascularized lymph node transfer (VLNT) and/or lympho-venous anastomosis (LVA) combined with peri-operative CPT
- Measurement outcome: relative interlimb volume difference (RID) (%) between the affected and non-affected side for the whole arm length and for segments of the arm (hand and wrist, lower arm and upper arm segment)
- Measurement instruments and moments: the Perometer® was used to measure arm volumes and to calculate volumes of the arm segments at baseline (T0) prior to intervention and 1 year following intervention (T1).

Results: 55 BCRL patients were included in three groups: LVA-group (N=35), VLNT-group (N=6) and LVA+VLNT-group (N=14). The patients had a mean age of 55,94 years old and a mean BMI of 25,4 kg/m² (SD ±4,3). In the first post-operative year, 93,9% of patients had manual lymphatic drainage; 77,6% wore a compression garment; 55,1% did exercise therapy and 100% got instructions for skin care.
Regarding the total group, there was a significant decrease in RID for the total arm between T0-T1 (p = .005). A significant decrease in RID was found for the hand-wrist segment in the LVA+VLNT group (p = .021). There was no significant change in RID for the lower arm segment in any of the groups. A significant decrease in RID for the upper arm segment was found in the total group (p = .034) and for the VLNT group (p = .035) (Table 1).
Conclusion: Microsurgical treatment of BCRL, combined with CPT may be an effective treatment method in order to prevent further arm volume increase and to help reducing arm volume. When conservative modalities are insufficient for the treatment of BCRL, microsurgery combined with CPT may reach this goal. Further research is necessary.

		p-value	Δ RID (%)
TOTAL group (N=55)	total arm	.005*	4.26
	wrist/hand	p>.05	3.32
	lower arm	p>.05	1.1
	upper arm	.034*	3.59
LVA-group (N=35)	total arm	p>.05	2.01
	wrist/hand	p>.05	-0.71
	lower arm	p>.05	0.45
	upper arm	p>.05	3.72
VLNT-group (N=6)	total arm	p>.05	3.23
	wrist/hand	p>.05	7.70
	lower arm	p>.05	1.11
	upper arm	.035*	4.48
LVA + VLNT-group (N=14)	total arm	p>.05	15.29
	wrist/hand	.021*	14.93
	lower arm	p>.05	3.67
	upper arm	p>.05	1.7

*significance p<.05
Table 1: arm (segment) volume changes between T_0 and T_1

LONG TERM OUTCOME OF LYMPHA TECHNIQUE AFTER 10 YEARS' FOLLOW-UP

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Purpose: To retrospectively assess the long term efficacy of the Lymphatic Microsurgical Preventive Healing Approach (LYMPHA) to prevent lymphedema after axillary dissection (AD) for breast cancer treatment.

Methods: Among 315 women referred from March 2008 to September 2020 to undergo complete AD plus LYMPHA technique for the prevention of arm lymphedema, 290 were followed up clinically up to 10 years by volumetry. All patients underwent preoperative lymphoscintigraphy (LS). Postoperatively, LS was performed after 5 and 10 years. Arm volume and LS alterations were assessed.

Results: Lymphedema rate was 2.5% (5/219) after AD plus LYMPHA plus Radiotherapy (219 women), 1.5% (1/71) after AD plus LYMPHA (71 women). No statistically significant differences in the arm volume were observed during follow-up in 284 patients and increase in volume in 6. Lymphoscintigraphy could demonstrate a stable lymphatic transport index (LTI) during follow-up in all patients except in 6, in whom the LTI increased.

Conclusions: LYMPHA represents a valid technique for primary prevention of secondary arm lymphedema and the outcome remains stable with time even after 10 years.

PROS AND CONS OF THE DIFFERENT MICROSURGICAL TECHNIQUES PROPOSED TODAY FOR THE TREATMENT OF LYMPHEDEMA

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Background: Different surgical techniques are nowadays proposed for the treatment of lymphedema. Most of them are reconstructive microsurgical techniques and some of them are debulking, modernly performed through mininvasive procedures. There is still not a consensus on which are the specific indications for each technique, how to evaluate results, pros and cons. The aim is to give an outline of the state of the art of the surgical treatment of lymphedema.

Materials & Methods: Various review articles and published clinical experiences from different groups of surgeons devoted to lymphatic surgery report about the usefulness of lymphatic reconstructive microsurgery in early stages, where there are less fibrotic tissue and lymphatic changes. It is much more difficult to find good lymphatics in more advanced stages, because of the severe sclerotic changes of the tissues and the aponeurotic fasciae, even though searching for both superficial and deep lymphatic collectors, actually a few number of lymphatic vessels may be found. It is mandatory to perform a proper preoperative diagnostic assessment through superficial and deep lymphoscintigraphy and echo-duplex venous scan. Lymphangio-MR is used in the most advanced stages to study the morphology of lymphatic drainage. ICG lymphangiography is used peroperatively together with the blue dye (BPV) to find sound lymphatic collectors during surgery. Results are evaluated by clinical assessment and lymphoscintigraphy. Minimally performed debulking techniques are used in the most advanced cases to reduce the fibrotic tissue after CDP plus microsurgery. Finally, microsurgical reconstructive techniques are used to prevent secondary lymphedema by performing lymphatic-venous anastomoses at the time of lymph nodal dissection (LYMPHA technique).

Results: The best results are obtained by microsurgery in the earlier stages and in secondary lymphedemas in terms of reduction of lymphangites, and less necessity of postoperative physical treatment and elastic garments. Microsurgery may be of great help also in late stages which may however need proper postoperative treatment to improve the outcome. Excellent results are obtained by LYMPHA technique in the prevention of secondary lymphedema following axillary and groin lymph nodal dissections.

Conclusions: The role of surgery in lymphedema treatment is of utmost importance mainly for secondary lymphedemas and obstructive primary ones. The meaning of microsurgical techniques is to open the tap and allow the lymph to pass through a lymphatic-venous by-pass as it happens in arterial surgery and in both cases the vein is the queen.

LIPOSUCTION NORMALIZES LYMPHEDEMA INDUCED ADIPOSE TISSUE HYPERTROPHY IN ELEPHANTIASIS OF THE LEG – HOW LONG DO OUTCOMES LAST?

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Introduction: Patients with long-standing pronounced non-pitting lymphedema do not respond to conservative treatment or microsurgical procedures because slow or absent lymph flow, as well as chronic inflammation, cause the formation of excess subcutaneous adipose tissue, which cannot be removed by these methods.

Objective: The swelling of chronic non-pitting arm lymphedema following breast cancer can be completely reduced by liposuction and has not recurred during 24 years' follow-up. Encouraged by this experience, we decided to evaluate the effectiveness of liposuction on leg lymphedema.

Material & Methods: 111 patients with a mean±SEM age of 49±1.5 years, and with a duration of leg swelling of 14±1.1 years underwent liposuction due to non-pitting, chronic lymphedema. There were 50 primary (PL), and 46 secondary lymphedemas (SL) following cancer therapy. Age at cancer treatment, and interval between cancer treatment and lymphedema start were 41±2.0 years and 2.5±0.7 years respectively. Age at onset of PL was 28±2.1 years (Figure 1, 2)

Results: Aspirate volume was 3642±167 ml with an adipose tissue concentration of 93±1.1% in the tourniquet fraction. Preoperative excess volume was 3733±188 ml. Postoperative mean reduction was 79±2.5% at 3 months and 100±2.3% at 1 year, and more than 100% during 15 years' follow-up, i.e. the lymphedematous leg was somewhat smaller than the healthy one. The preoperative mean ratio between the volumes of the edematous and healthy legs was 1.4±0.02, rapidly declining to 1.0±0.01 at 1 year and less than 1 after one year.

Conclusions: Liposuction is an effective method for treatment of chronic, non-pitting leg lymphedema in patients who have failed conservative treatment. It is the only known method that completely reduces excess volume at all stages of lymphedema. The removal of hypertrophied adipose tissue is a prerequisite to complete reduction. The reduced volume is maintained through constant use of compression garments.

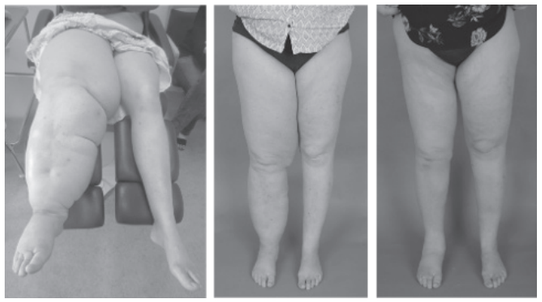

Fig. 1. A 45-years-old woman with a heavy pitting secondary leg lymphedema of 25 650 ml since 8 years following treatment of a cervix cancer (left). After conservative treatment the lymphedema was reduced to 5 850 ml before liposuction. Postoperative result 3 years after liposuction with complete reduction (right).

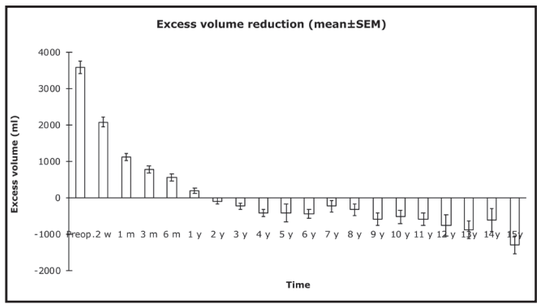

Fig. 2. Mean pre-and postoperative excess volume reduction following liposuction of leg lymphedema.

FROM LYMPH TO FAT – LESSONS LEARNED DURING 25 YEARS OF COMPLETE REDUCTION OF ARM LYMPHEDEMA

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Introduction: Patients with chronic non-pitting lymphedema do not respond to conservative treatment because diminished lymph flow and inflammation result in the deposition of excess adipose tissue subcutaneously. Microsurgical reconstructions, although attractive as a physiological concept, cannot provide complete reduction in chronic non-pitting lymphedema because they do not eliminate the newly formed subcutaneous adipose tissue collections. To remove the excess adipose seems thus to be a logical treatment strategy.

Objective: This prospective study describes the long-term outcome of liposuction of arm lymphedema.

Material & Methods: 182 women with non-pitting edema with a mean±SEM age of 63±0.8 years, and with a mean duration of arm swelling of 8.7±0.5 years underwent liposuction. Mean age at breast cancer operation, mean interval between breast cancer operation and lymphedema start, and duration of lymphedema were 51±0.8 years, 2.7±0.4 years, and 8.7±0.5 years respectively. Aspirate and arm volumes were recorded.

Results: Aspirate mean volume was 1768±49 ml with an adipose tissue concentration of 95±0.8 % in the tourniquet fraction. Preoperative mean excess volume was 1492±57 ml. Postoperative mean reduction was 103±2.1 % at 3 months and 118±28% at 1 year, and more than 100% during 24 years' follow-up, i.e. the lymphedematous arm was somewhat smaller than the healthy arm. The preoperative mean ratio between the volumes of the edematous and healthy arms was 1.5±0.02, rapidly declining to 1.0±0.01 at 3 months, and less than 1 after 6 months up to 25 years after surgery.

Conclusions: Liposuction is an effective method for treatment of chronic, non-pitting arm lymphedema in patients who have failed conservative treatment. Because of adipose tissue hypertrophy, it is the only known method that completely reduces excess volume at all stages of arm lymphedema. Removing the hypertrophied adipose tissue is a prerequisite to achieve complete reduction. The newly reduced volume is maintained through constant use of compression garments.

Fig. 1. A 57-years-old woman with a non-pitting secondary lymphedema of 4 235 ml since 5 years following breast cancer treatment. Complete reduction 6 months after liposuction.

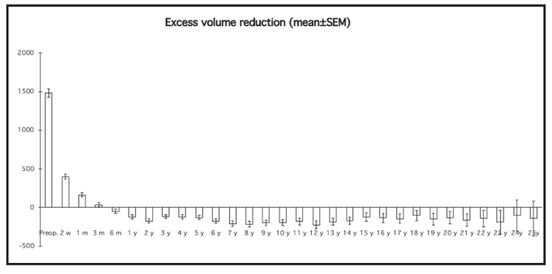

Fig. 2. Mean pre-and postoperative excess volume reduction following liposuction of arm lymphedema.

STAGING-GUIDED AND LONG-LASTING SURGICAL TREATMENT OF PERIPHERAL LYMPHEDEMA – THE GALT SYSTEM (GENOA ADVANCED LYMPHEDEMA THERAPY)

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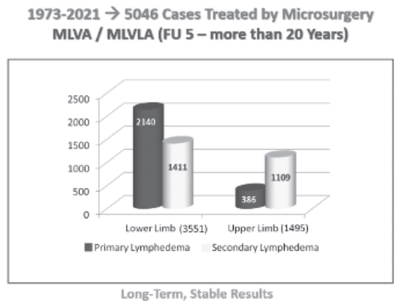
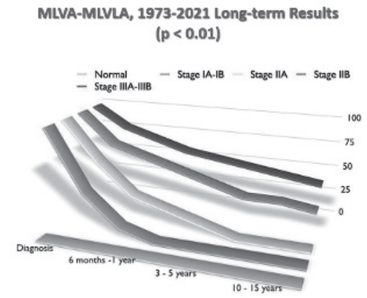
Background: Peripheral lymphedema remains a poorly recognized disease that causes significant morbidity and chronic lymphedema is associated with fibro-adipose tissue changes ('non-pitting' edema) that is irreversible when untreated. **Lymphatic Microsurgery provides a functional repair to overcome the obstacle in lymphatic flow. In these cases, Lymphatic Microsurgery helps to resolve the lymphstasis that contributes markedly to swelling. Notwithstanding the success of the microsurgery, there often remains significant fibro-adipose tissue in the affected limb in advanced lymphedema, which contributes to residual lymphstasis and increased risk of infection. The Authors underline a recently developed Fibro-Lipo-Lymph-Aspiration technique to improve this chronic swelling, using a Lymph Vessel Sparing Procedure (FLLA-LVSP).**

Methods: For the preliminary Lymphatic Microsurgery, the lymphatic collectors (superficial and deep) are anastomosed with telescopic technique to multiple tributary veins in a single-site procedure. **In 500 cases of advanced lymphedema, we used a recently developed, complementary and sequential, minimally invasive surgical approach FLLA-LVSP, where Indocyanine Green Fluorescent Microlymphography highlight the superficial lymphatic pathways and the excess fibro-adipose tissue is carefully aspirated.**

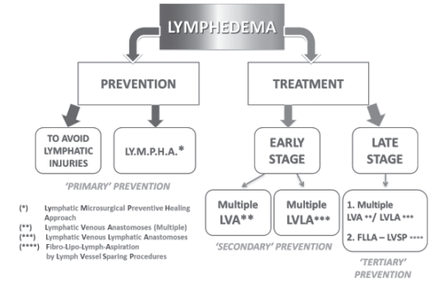
Results:

1. Lymphatic Microsurgery – MLVA / MLVLA:





2. Fibro-Lipo-Lymph Aspiration with Lymph Vessel Sparing Procedure (FLLA-LVSP):
   For the advanced cases involving the upper limb, there was an average pre-surgery excess volume of 24.16%, which reduced to 2.76% after the FLLA-LVSP (Z-score =-6.73, p<0.001). Similarly, for the lower limb, there was an average pre-surgery excess limb volume of 22.84% and a reduction to 2.84% post-operatively (Z-score=-3.42, p<0.01).
3. Genoa Algoritm for Lymphedema Surgery:



MICROLYMPHATIC PUMPS FOR LONGSTANDING LYMPHEDEMA

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Longstanding lymphedema have the disadvantage that the propulsion of lymph within the main collectors is disturbed due to their fibrotic wall. However always some lumen is remaining open.

Suction of lymph at the cranial end of the collectors would help to transport lymph along the main collectors. It was proved that isolated lymphatic collectors are able to continue their pumping activity and to suct lymph out of a reservoir.

Independent investigators report about an astonishing effect of lymphatic vessel grafts in longstanding lymphedema.

The ability of lymphatic vessel grafts to act as a suction pump may explain this effect.

This is furthermore elucidated by scintigraphic studies. In longstanding lymphedema often no collectors are visible preoperatively. After the grafting with time functioning lymphatic vessels become visible together with the reduction or dissappearance of sings of peripheral lymph congestion.

The experimental and clinical data together with clincial and scintigraphic examples are discussed in detail.

Conclusions: MLVA techniques when performed at a single-site produce excellent outcomes in the treatment of lymphedema, giving the possibility of complete restoration of lymphatic flow in the Early Stages, when tissue changes are minimal. In cases of Advanced Lymphedema, the FLLA-LVSP is significantly effective from not only the clinical and functional point of view, but with immediate cosmetic results too. More importantly, the removal of excess tissue is completed without further damage to lymphatic vessels.

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LONG-TERM EFFECTS OF SURGICAL TREATMENT OF SECONDARY LYMPHOEDEMA. DO I STILL NEED COMPLEX DECONGESTIVE THERAPY AFTERWARDS?

PROF. CORNELY M.

Secondary lymphoedema of an extremity leads to an increase in the volume of the extremity with a consequent increase in circumference. From 2005 to 2020, 46 patients with secondary lymphoedema after breast cancer and gynaecological tumours were treated in our practice by means of resective surgery. We use a special surgical procedure called “Lymphological Liposculpture”, a two-pronged approach of surgery and conservative follow-up treatment to achieve curative and sustainable volume and circumference equalization of the extremities.

This procedure also reduces the need for complex decongestive therapy (CPD) by up to 80% and more. Questionnaires were used to assess changes in quality of life and the continued use of conservative measures, and perometry was used to measure postoperative volume equalization of the extremities. All data were evaluated as long-term results up to 15 years after “Lymphological Liposculpture”.

The alignment of the affected side with the healthy side was achieved in all patients, and movement restrictions caused by the lymphoedema were significantly improved.

96% of the patients described a permanent reduction in conservative treatments with compression and lymphatic drainage (CPD). In 56% of the 46 women who were followed up, this CPD treatment could even be stopped completely postoperatively. 87% experienced a significant improvement in their quality of life,

We also recommend resection of the subcutaneous matrix in cases of long-standing secondary lymphoedema according to the standardised curative and sustainable procedure “Lymphological Liposculpture” for circumferential adjustment of the extremities, consecutive reduction of CPE and improvement of the quality of life.

LONG TERM PATENCY OF MLVA

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Aim: The aim of the study is to assess the long term patency of the technique of multiple lymphatic-venous anastomosis (MLVA) to treat peripheral lymphedema.

Material & Methods: From January 2014 to December 2014, 150 patients affected from limb lymphedema were treated by microsurgery using MLVA. 85 had lower limb lymphedema (47 primary and 38 secondary), 65 upper limb (63 secondary and 2 primary). Secondary lower limb lymphedemas followed uterine cancer (14), prostate cancer (11), ovarian cancer (5), and trunk skin melanoma (8). Secondary upper limb lymphedema were all related to breast cancer treatment (63). All lower and upper primary lymphedemas were due to truncular lymphatic malformation and specifically to lymph nodal malformation (LAD II, according to Papendieck’s classification). As regards the staging (following ISL staging) of the disease, 47 lower lymphedema (25 primary and 22 secondary) were at early stage 2, and 38 at late stage 2 (22 primary, 16 secondary). 36 secondary upper limb lymphedemas were at early stage 2 and 27 late stage 2. The two primary upper limb lymphedema were at early stage 2. All patients had combined decongestive physiotherapy (CDP) for 6 months at least and all of them had improvement of the disease in terms of stage reduction (from stage 3 to late stage 2, and from late stage 2 to early stage 2). All of them did not have a stability of the result obtained with the intensive phase of the conservative treatment notwithstanding a proper compression garment and the maintenance phase (once a month combined treatment). 77% (116 patients, 65 lower – 36 primary and 29 secondary – and 51 upper – 50 secondary and 1 primary – limb lymphedemas) had 1 to 3 episodes of acute lymphangitis per year. All of them had lymphoscintigraphy (superficial and deep), venous echo-Doppler and abdominal (for lower limbs) or axillary (for upper limbs) echography preoperatively. Primary and secondary lower limb lymphedemas were surgically treated by MLVA (3-6 lymphatics dunked into one vein) performed just under the groin crease after injecting the blue dye (Blue Patent V – BPV) and indocyanine green (ICG) 15 cm downstream at 4 sites from the internal to the anterior side, into the dermal, subcutaneous and perifascial areas. Results were assessed by limb volume (by the truncated cone formula) taken preop and at 1, 3 and 5 years postop, lymphoscintigraphy (measuring lymphatic transport index – TI – normal value < 10) performed preoperatively and after 12 and 24 months from operation, and number of episodes of lymphangitis per year. MLVA patency was assessed by the lymphatic TI, based particularly on the tracer transit kinetic, the distributive pattern (dermal back flow – DBF) and the visualization of lymphatics, whilst the other two parameters – lymph node uptake and visualization time – were substituted with the disappearance of the tracer at the site of MLVA, due to the passage of lymph into the blood stream and the liver uptake time.

Results: Early stage 2 upper (36) and lower (22) limb secondary lymphedemas had 75-90% excess volume reduction and early stage 2 upper (2) and lower (25) limb primary lymphedemas 60-75%. Late stage 2 upper (27) and lower (15) limb secondary lymphedemas and lower limb primary lymphedemas (19) had 45-60% excess volume reduction. One late stage 2 lower limb lymphedema and 3 late stage primary lower limb lymphedema had only 30% excess volume reduction. The volume improvement was already found at 1 year postop and remained stable in the 5-year follow-up. Lymphatic TI showed an improvement from 25-37 preoperatively to 8-14 after 12 and 24 months from MLVA and together with the disappearance of the tracer at the site of the anastomosis and the earlier liver uptake showed the MLVA patency in 146 patients. It was not possible to demonstrate MLVA patency in the 4 patients (1 late stage 2 secondary lower limb and 3 late stage 2 primary lower limb lymphedemas), those who had only 30% excess volume reduction. All patients had no lymphangites postoperatively.

Conclusions: This study allowed to demonstrate the patency of MLVA for the treatment of primary and secondary upper and lower limb lymphedemas in 97% of the patients. Moreover, better results were obtained in secondary lymphedemas of extremities, above all in upper limbs, and at earlier stages.

SURGICAL TREATMENT OF POSTMASTECTOMY IN ADVANCED LYMPHEDEMA THROUGH THE SUPER CHARGE SUBMENTAL LYMPHATIC TRANSFER

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Background: The purpose of this study was to evaluate the outcome of vascularized submental lymph node transfer using a super-charge technique in patients with advanced (grade III/III) secondary upper limb lymphedema developed after mastectomy and homolateral lymph node resection.

Material & Methods: Between December of 2014 and August of 2019, 8 consecutive patients underwent a vascularized submental supercharge bilateral lymph node transfer. All patients were operated at the Universital Hospital of Alicante by the same surgeon with a super-charge technique, doing multiple vascular anastomosis in the proximal third of the forearm. We motorized the reductions of operated arm at 1, 3, 6, 12 months and we registered the incidence of cellulitis. Moreover, we dispensed a validated survey of the quality of life specific for the upper limb lymphedema (ULL-27).

Results: Among these, 8 female patients with a mean age of 58,6 years (-10,6/+16,4) and a median body mass index (BMI) of 25,6 (-4,6/+6,4) kg/m2 we found that the mean reduction of the arm circumference was 12,7% (-11,2/+30,9) at 1 month, 15% (-14,1/+28,3) at 3, 7,7% (-23,8/+47,7) at 6, 13,1%(-5,9/+56,4) at 12 months at a mean follow up of 39,3 (-29,3/+70,7) months. According to the survey ULL-27 we pointed out a rate in terms of quality of life of the patients after the surgery of 25,6 points (-18.4/+ 32.6).

Conclusions: Vascularized submental lymph node transfer using the super-charge technique is a novel and reliable procedure that significantly improves not only the secondary advanced lymphedema of the upper extremity, but also has a positive impact on the patients quality of life.

MICROSURGICAL TREATMENT OF LYMPHEDEMA

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Microsurgical treatment of lymphedema aims to improve the lymphatic drainage function to reduce lymph stasis and achieve a better local immunity.

Although decongestive therapy and compressive garment are the mainstay of treatment, there is sufficient evidence in the literature that microsurgery can reduce patients' symptoms and improve quality of life.

Correct indication and precise technique are mandatory to achieve good results, improving the efficacy of physical therapy and of compression, and in a few early stages, to discontinue conservative therapies for a long time.

The two main microsurgical procedures are lymphatic-venous anastomosis (LVA) and vascularized lymph node transfer (VLNT).

The prerequisite for the efficacy of LVA is to operate on lymphatic vessels free of sclerotic changes, that still have a good peristaltic function, carrying the lymph into a venous circulation without blood backflow.

VLNT technique transfers a lymph node flap with its vascular pedicle into the limb affected by lymphedema, with a microvascular anastomosis to the limb recipient vessels.

After neolymphangiogenesis occurs, the connections between the lymphatics of the limb and the flap allows to direct the lymph into the vein of the flap, through the natural lymphatic-venous anastomosis present in the lymph nodes.

The author describes the indications, technical tricks and preoperative planning to maximize the results and select patients for surgery.

In the selected cases candidate for surgery, patients' expectations must be assessed according to the severity of disease, with the best results typically achieved in the early stages.