THE MANAGEMENT OF LYMPHOSTATIC FIBROSIS

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KLOSE LYMPHEDEMA CONFERENCE 2019

FIBROSIS IMPACTS OUR PATIENTS:

All lymphedema patients have fibrosis!

WHY IS FIBROSIS IMPORTANT?

- FIBROSIS = DAMAGE TO LYMPHATIC SYSTEM
- TREATING FIBROSIS INFLUENCES CURRENT LYMPHEDEMA AND LONG TERM PROGNOSIS
- EARLY TREATMENT BEST LONG TERM OUTCOMES
- TOOLBOX APPROACH- MANY OPTIONS
- COI DISCLOSURES

WHAT IS FIBROSIS?

- ABNORMAL THICKENING OF TISSUE
- EXCESS FIBROUS CONNECTIVE TISSUE
- PRESENT IN ALL STAGES OF LYMPHEDEMA

- TWO BASIC TYPES:
  - SOFT, THICKENED TISSUE: FAT
  - HARDENED TISSUE: SCAR

GOALS FOR TREATING FIBROSIS

- LESSEN SYMPTOMS OF LYMPHEDEMA AND FIBROSIS: PAIN, IMMOBILITY, DYSFUNCTION
- REPAIR OR COMPENSATE FOR FIBROTIC DAMAGE TO THE LYMPHATIC SYSTEM
- INFLUENCE FIBROSIS FORMATION/MATURATION
- INFLUENCE FUTURE PROGRESSION OF LYMPHEDEMA

FIBROSIS ETIOLOGY

- FIBROSCLEROSIS: NORMAL HEALTHY TISSUE REPLACED BY SCAR TISSUE AND/OR FATTY TISSUE
- CREATES ALTERED TISSUE COMPOSITION AND DENSITY
- CAN CREATE CHRONIC CONGESTION BY IMPEDING LYMPHATIC CIRCULATION

TYPES OF FIBROSIS:

SOFT:

- LYMPHOSTATIC FIBROSIS FROM LYMPH STASIS

HARD:

- SURGICAL SCAR TISSUE (SUPERFICIAL AND DEEP)
- RADIATION INDUCED FIBROSIS
- POST-CELLULITIS FIBROSIS
- CORDING (AXILLARY WEB SYNDROME)

LYMPHOSTATIC FIBROSIS

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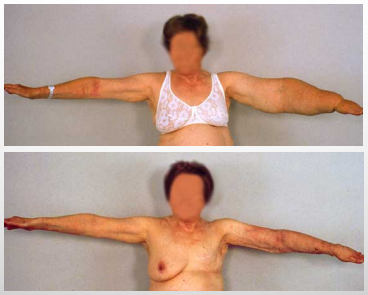
- PRESENT IN ALL STAGES OF LYMPHEDEMA!
- CREATED FROM LYMPH STASIS
- ACUTE STAGE: EDEMA THICKENS TO "GEL"
- CHRONIC STAGE: HARDER AND THICKER OVER TIME
- "WOODY" FIBROSIS FREQUENTLY OCCURS AT TOES AND MALLEOLI
- BRORSON: SUBCUTANEOUS FIBROSIS EXTRACTED LIPOSUCTION: 10% THICKENED LYMPH, 90% FAT

LYMPHOSTATIC FIBROSIS AND LIPOSUCTION



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LYMPHOSTATIC FIBROSIS

- LYMPHOSTASIS > HIGH PROTEIN EDEMA > ACCUMULATION OF IMMUNE CELLS > FIBROSCLEROSIS> FAT DEPOSITION
- LYMPHOSTATIC FIBROSIS CAN OCCUR 2° TO OTHER FIBROSIS (SURGERY, RADIATION, CELLULITIS), AND INJURY AND DISEASE FACTORS
- CURRENT RESEARCH: *INFLAMMATORY PROCESS* TRIGGERS LYMPHOSTATIC FIBROSIS

STAGES OF LYMPHEDEMA (FÖLDI):

STAGE 0 (LATENCY):

- FOCAL **FIBROSCLEROTIC** TISSUE ALTERATIONS

STAGE I (REVERSIBLE):

- HIGH PROTEIN EDEMA,
- FOCAL **FIBROSCLEROTIC** TISSUE ALTERATIONS

STAGE II (SPONTANEOUSLY IRREVERSIBLE) AND STAGE III (ELEPHANTIASIS):

- EXTENSIVE **FIBROSCLEROSIS,**
- PROLIFERATION OF **ADIPOSE** TISSUE

LYMPHOSTATIC FIBROSIS STAGE O (LATENCY):

FOCAL FIBROSCLEROTIC TISSUE ALTERATIONS

LYMPHOSTATIC FIBROSIS STAGE O (LATENCY):

FOCAL FIBROSCLEROTIC TISSUE ALTERATIONS

LYMPHOSTATIC FIBROSIS STAGE I (REVERSIBLE):

HIGH PROTEIN EDEMA

FOCAL FIBROSCLEROTIC TISSUE ALTERATIONS

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LYMPHOSTATIC FIBROSIS STAGE II (IRREVERSIBLE):

EXTENSIVE FIBROSCLEROSIS

PROLIFERATION OF ADIPOSE TISSUE

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EXTENSIVE FIBROSCLEROSIS

PROLIFERATION OF ADIPOSE TISSUE

LYMPHOSTATIC FIBROSIS STAGE III ELEPHANTIASIS:

EXTENSIVE FIBROSCLEROSIS

PROLIFERATION OF ADIPOSE TISSUE

LYMPHOSTATIC FIBROSIS STAGE III ELEPHANTIASIS:

EXTENSIVE FIBROSCLEROSIS

PROLIFERATION OF ADIPOSE TISSUE

TYPES OF SOFT TISSUE FIBROSIS:

SOFT:

- LYMPHOSTATIC FIBROSIS FROM LYMPH STASIS

HARD:

- SURGICAL SCAR TISSUE (SUPERFICIAL AND DEEP)
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- CORDING (AXILLARY WEB SYNDROME)

SURGICAL FIBROSIS

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- WOUND HEALING OCCURS FOR THREE WEEKS BEFORE SCAR TISSUE FORMATION BEGINS
- AFTER THREE WEEKS COLLAGEN FIBERS BEGIN TO CROSS-LINK AND FORM THE SCAR MATRIX
- TYPICAL MATURATION TIME FOR SCAR TISSUE CAN BE UP TO TWO YEARS
- SURGICAL SCAR TISSUE IS BOTH SUPERFICIAL AND DEEP
- DEEP SCARRING CAN AND DOES OCCUR IN DIFFERENT LOCATIONS THAN THE SUPERFICIAL SCAR

SURGICAL FIBROSIS: INFLUENCING FACTORS

- INFLAMMATION
- CO-MORBIDITIES: DIABETES, CIRCULATORY
- KELOID FORMATION
- COMPLICATIONS: EDEMA, CELLULITIS, NECROSIS
- CHEMOTHERAPY (PRE OR POST)
- RADIATION THERAPY (PRE OR POST)
- TOPICAL SILICONE DRESSINGS/OINTMENT
- 6 MOS POST-OPERATIVE WINDOW OF OPPORTUNITY

SURGICAL FIBROSIS: INFLUENCING FACTORS

- TYPE OF SURGERY
- SKILL AND SPECIALTY OF SURGEON (PLASTIC VS. GENERAL)
- PRIOR SCAR TISSUE: MULTIPLE SURGERIES AT SAME SITE
- DEGREE OF POST-SURGICAL SWELLING
- PRIOR LYMPHEDEMA
- AMOUNT OF TISSUE AND LYMPH NODES EXCISED
- TISSUE CONSERVATION LEFT FOR FUTURE RECONSTRUCTION
- TYPES OF INTERNAL AND EXTERNAL FASTENING AGENTS: STAPLES, GLUE, VARIOUS TYPES OF SUTURES, STERI STRIPS

RADIATION-INDUCED FIBROSIS

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- ALTERED TISSUE COMPOSITION >THINNER, HARDER AND MORE BRITTLE
- MECHANICAL FACTORS INFLUENCING RADIATION DAMAGE: TYPE, DURATION, INTENSITY, AMOUNT AND LOCATION OF THE RADIATION FIELD
- PATIENT-ORIENTED FACTORS: SKIN SENSITIVITY AND TYPE, TISSUE DENSITY AND COMPOSITION
- LONG-TERM EFFECTS: HEART DISEASE, OSTEOPOROSIS, TOOTH LOSS

POST-CELLULITIS FIBROSIS

POST-CELLULITIS FIBROSIS

- ACUTE, INTENSE CONGESTION OF FLUID
- HIGH DENSITY OF SOLIDS INCLUDING INFECTIOUS WASTES
- OFTEN FORMS A DENSE AND WOODY AREA AT AND AROUND INFECTION SITE
- SOME PATIENTS ARE VULNERABLE TO MULTIPLE CELLULITIS REOCCURRENCES AT THE SAME OR DIFFERENT SITES

CELLULITIS: INFLUENCING FACTORS

- DEPRESSED IMMUNE SYSTEM (CHEMOTHERAPY)
- DIABETES
- CIRCULATORY DISORDERS
- GASTRIC INFLAMMATORY FACTORS:
  - FOOD SENSITIVITIES
  - CELIAC DISEASE
  - GLUTEN INTOLERANCE- PRIMARY LYMPHEDEMA

CORDING

CORDING

- ALSO CALLED AWS, OR AXILLARY WEB SYNDROME
- VISIBLE AND PALPABLE CORD OF TISSUE CAN CREATE DISCOMFORT AND FUNCTIONAL RESTRICTIONS
- TYPICALLY BETWEEN AXILLA AND ANTECUBITAL FOSSA CAN AFFECT ENTIRE UPPER QUADRANT INCLUDING TRUNK AND HAND
- THROMBOSIS OF LYMPHATIC OR VENOUS VESSELS, OR LYMPHATIC FIBROSIS

FIBROSIS CAN BE AS BIG OF A PROBLEM AS LYMPHEDEMA

FIBROSIS-ASSOCIATED IMPAIRMENTS

- FASCIA CHANGES
- TISSUE CONGESTION: THICKENED TISSUE IMPEDES LYMPHATIC CIRCULATION
- CAN LEAD TO VICIOUS CYCLE INCREASING FIBROSIS
- IMPAIRED COSMESIS (BODY IMAGE)
- INCREASED TISSUE DENSITY (WEIGHT AND BULK)
- DERMAL CHANGES (PUCKERING, LUMPINESS)
- INCREASED RISK OF INFECTION

FIBROSIS-ASSOCIATED IMPAIRMENTS

FUNCTIONAL IMPAIRMENTS:

- INCREASED WEIGHT AND BULK AND ASYMMETRY CAN CREATE POSTURAL IMBALANCES
- DECREASED MOBILITY
- DECREASED SELF CARE ABILITY
- INCREASE FALL RISK

PAIN: SECONDARY TO SOFT TISSUE/NERVE IMPINGEMENT EITHER BY FIBROTIC TISSUE OR LYMPHEDEMA SECONDARY TO FIBROTIC TISSUE

MEASURING FIBROSIS

PALPATION:

- LOCATION, IDENTIFICATION AND MEASUREMENT OF AREA (DON'T FORGET THE BACK!)
- DEPTH: SUPERFICIAL VS. REGIONAL
- QUALITY OF TISSUE: "GEL" VS. "WOODY"
- QUANTIFY BY SUBJECTIVE THERAPIST ASSESSMENT: "20% OF NORMAL TISSUE EXTENSIBILITY"

MEASURING FIBROSIS

SCARS:

- ADHEREMETER (DEGREE SCAR ADHERENCE)
- CUTOMETER (ELASTICITY)
- TISSUE ULTRASOUND PALPATION SYSTEM (TUPS)
- VANCOUVER SCAR SCALE (VSS)
- PATIENT AND OBSERVER SCAR ASSESSMENT SCALE (POSAS)

MEASURING FIBROSIS

TOOLS:

- MEASUREMENT RESISTANCE OF TISSUES TO INDENTATION (TONOMETER, DELFIN ELASTIMETER)
- CALIPERS
- BIOIMPEDANCE/BODY COMPOSITION: COMPARE FAT MASS VS. FAT FREE MASS
- ULTRASOUND IMAGING

DOCUMENTING FIBROSIS

- TYPE
- ASSOCIATED EDEMA
- LOCATION(S)
- PAIN W/ AND W/O MOVEMENT OR ACTIVITY
- TISSUE QUALITY I.E.: WOODY, SPONGY
- PALPABLE RESTRICTION OF TISSUE EXPRESSED IN PERCENTAGE: "**R AXILLA TISSUE EXTENSIBILITY IMPROVED FROM 5% TO 15% OF NORMAL.**"

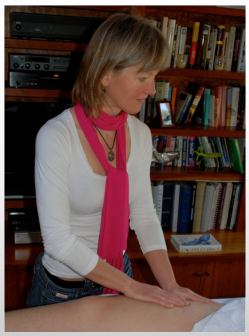
TREATING FIBROSIS

- ADDRESS SWELLING FIRST!
- BE AWARE OF CONTRAINDICATIONS
- START CAUTIOUSLY, MONITOR ADVERSE SX
- RISK REDUCTION TO LOWER BODY MASS AND INFLAMMATION ARE IMPORTANT
- EMPOWER PATIENT: EDUCATION AND TOOLS
- EVEN LONG-STANDING, CHRONIC FIBROSIS CAN CHANGE

TREATMENTS FOR FIBROSIS

- MANUAL THERAPY
- PNEUMATIC COMPRESSION
- BANDAGING AND ANTI-FIBROTIC GARMENTS
- MODALITIES

MANUAL THERAPY



MANUAL THERAPY

- ANTI-FIBROTIC MASSAGE
- MFR (MYOFASCIAL RELEASE)
- TRIGGER POINT AND BALL ROLLING MASSAGE
- INSTRUMENT ASSISTED SOFT TISSUE MOBILIZATION (IASTM, FASCIA BLASTER)
- A/AA/PROM WITH/WITHOUT RELEASE
- NEURAL TENSION TECHNIQUES
- DYCEM/GLOVES: INCREASED SKIN TRACTION

MANUAL THERAPY PRECAUTIONS

OBSERVE ALL STANDARD MLD PRECAUTIONS

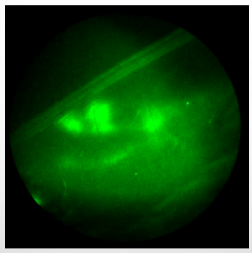
TAKE EXTRA CARE WITH:

- PAIN
- ALTERED SENSATION (HYPERSENSATE, INSENSATE)
- FRAIL SKIN
- RECENT SURGERY, RADIATION, CHEMOTHERAPY

PNEUMATIC COMPRESSION

PNEUMATIC COMPRESSION

- PERFORMS BOTH DECONGESTION AND COMPRESSION
- HIGHER PRESSURES REMODEL FIBROSIS
- EFFECTIVE COMPONENT OF LYMPHEDEMA AND FIBROSIS TREATMENT IN CLINIC AND AT HOME
- INCREASES LYMPHATIC FUNCTION (ALDRICH 2016)



WHICH PATIENTS ARE APPROPRIATE FOR PNEUMATIC COMPRESSION?

SWELLING AND FIBROSIS CAN BE DIFFICULT FOR SOME PATIENTS TO SELF MANAGE

- LIMITED ROM, STRENGTH, DEXTERITY
- PAIN
- LACK OF CAREGIVER SUPPORT
- PROFOUND LYMPHATIC DAMAGE: FIBROSIS

MLD AND PNEUMATIC COMPRESSION

- MLD CAN BE USED ANYWHERE AND TAILORED FOR SPECIAL NEEDS
- PNEUMATIC COMPRESSION: MECHANIZED DECONGESTION DELIVERS PROGRAMMED TREATMENT
- CAN WORK TOGETHER: MLD CAN STIMULATE AREA NOT COVERED BY PUMP (I.E.: SUPRACLAVICULAR FOSSAE, AREAS SUCH AS AXILLA REQUIRING FOCUSED TREATMENT.

ADVANTAGES OF PNEUMATIC COMPRESSION

- ACCEPTANCE AND COMPLIANCE (RIDNER 2008)
- EASY TO USE (AVERY 2000)
- IMPROVES PHYSICAL FUNCTION (KARACA-MANDIC 2015, CAMEROTA 2011)
- RELIABLE, CONSISTENT TREATMENT (SZOLNOKY 2009, SZUBA 2002)
- DECREASED PAIN (LONIGAN 2016)
- INSURANCE COVERAGE (CMS 2002)

VERSATILE APPLIANCES

- BILATERAL OR UNILATERAL TREATMENT
- BASIC APPLIANCES TREAT EXTREMITIES
- APPLIANCES CAN ALSO TREAT ONE OR TWO EXTREMITIES AS WELL AS HEAD/NECK AND TORSO (CHEST, ABDOMEN, HIPS, GENITALS.)
- SPECIALIZED SYSTEMS FOR OBESE PATIENTS

TYPES OF APPLIANCES

- LEG SLEEVE
- ARM SLEEVE
- TRUNK + LEGS
- CHEST + ARM(S)
- HEAD/NECK + CHEST

PNEUMATIC COMPRESSION MYTHS

NOT AS EFFECTIVE AS MLD?

- BETTER MAINTENANCE EDEMA CONTROL THAN SELF-ADMINISTERED MLD (WILBURN 2006)
- MLD MAY NOT BE WELL-TOLERATED IN PATIENTS WITH WOUNDS (BETZ 2008)

MLD MOVES LYMPH, PNEUMATIC COMPRESSION MOVES FLUID AND LEAVES BEHIND PROTEINS?

- INDICATORS OF RESIDUAL PROTEINS (DECREASED ROM, CUTANEOUS FIBROSIS) NOT EVIDENT AFTER PERIOD OF PNEUMATIC COMPRESSION USE (SZUBA 2002)

HIGH RISK OF GENITAL EDEMA?

- BORIS 1998 STUDY: LEG SLEEVE APPLIANCES ONLY
- NO ABDOMINAL OR GENITAL TREATMENT
- MANY STUDY PATIENTS HAD PRE-EXISTING GENITAL LYMPHEDEMA
- CRITICAL FOR THERAPIST TO FOLLOW UP WITH PATIENTS WHO HAVE LEG SLEEVE APPLIANCES TO ASSESS FOR CUFFING AT PROXIMAL MARGIN OF SLEEVE AND/OR PERSISTENT PROXIMAL EDEMA.
- PANTSUIT APPLIANCE TREATS THE GENITAL AREA.

HIGH AMOUNTS OF COMPRESSION ALWAYS CAUSES TISSUE DAMAGE?

When standing in water at a depth of one meter distal compression is *75 mmHg* and pressure increases with movement.

PRESSURE IN RESEARCH

- **120** MMHG THE HIGHER THE COMPRESSION THE LARGER THE FLUID FLOW VOLUME (ZALESKA 2018)
- **>100** MMHG REQUIRED TO INFLUENCE SUBDERMAL PRESSURE IN FIBROTIC PATIENTS (OLSZEWSKY 2010)
- **160-180** MMHG (MANJULA 2002)

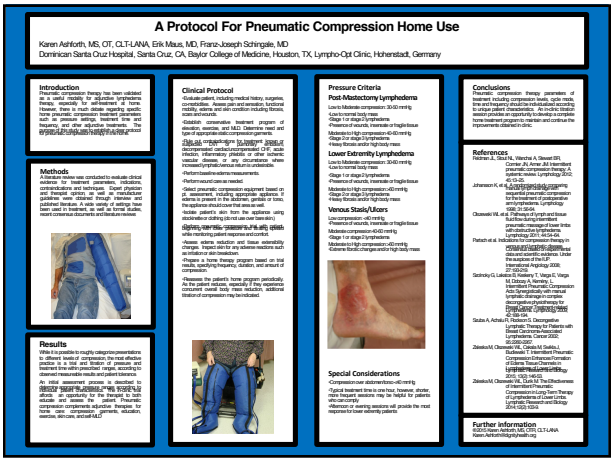
SELECTING TREATMENT PARAMETERS

TIME, PRESSURE AND FREQUENCY SHOULD BE SELECTED ACCORDING TO THE PATIENT'S INDIVIDUAL CHARACTERISTICS:

- EDEMA
- FIBROSIS
- WOUND STATUS
- BODY MASS
- RESPONSE TO TRIAL OF COMPRESSION

GUIDELINES FOR SETTING PRESSURE

- THICKER TISSUES NEED HIGHER PRESSURES
- PRESSURE MAY NEED TO BE INCREASED/LOWERED DEPENDING UPON CHANGES IN BODY MASS/FIBROSIS
- TITRATE PRESSURE TO PATIENT COMFORT
- QUILTED COMPRESSION ADDS "DUAL MASSAGE:" MAY INCREASE TOLERANCE FOR HIGHER PRESSURE
- REFER TO THE TREATMENT PROTOCOL FOR SPECIFIC GUIDELINES

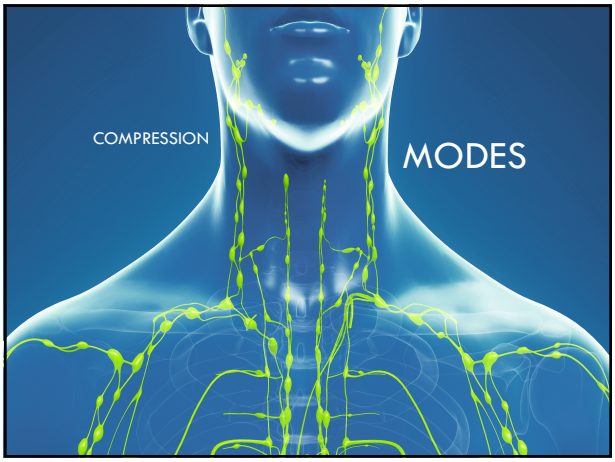


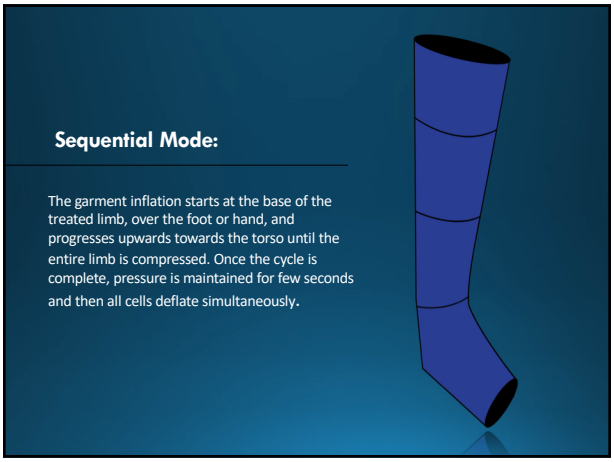
TREATMENT SESSION

- COVER TREATED AREA: STOCKINETTE, CLOTHING
- MAY USE PUMP GARMENT OVER COMPRESSION GARMENTS, BUT NEED TO MONITOR EFFECTS
- PERFORM TRIAL FOR 30 MINUTES, ADJUSTING PRESSURE PER PATIENT COMFORT
- AFTER TREATMENT, CHECK FOR EDEMA REDUCTION, SOFTENING AND TISSUE EXTENSIBILITY CHANGES
- EXAMINE SKIN TO ENSURE NO ADVERSE EFFECTS

HOME TREATMENT PROGRAM

- DETERMINE TREATMENT TIME AND FREQUENCY
- STANDARD TREATMENT: ONE HOUR DAILY
- SOME PATIENTS MAY BENEFIT FROM ADDITIONAL OR SHORTER SESSIONS SUCH AS 30-60 MINUTES, 1-2X DAY
- COMPRESSION GARMENTS MAINTAIN THE EDEMA REDUCTION





PNEUMATIC COMPRESSION SETTINGS

SEQUENTIAL PROGRAMMING:

- MORE AGGRESSIVE THAN PERISTALTIC
- SUSTAINED DISTAL PRESSURE VERY USEFUL FOR FIBROSIS ON HANDS AND FEET, FOREARMS AND LOWER LEGS
- PADDING FEET/HANDS WITH TOWELS OR QUILTED COMPRESSION GARMENTS MAY DISPERSE DISTAL PRESSURE AND INCREASE TOLERANCE FOR SEQUENTIAL MODE

Peristaltic Mode:

The garment inflation starts at the base of the limb, over the foot or hand. As the cell at the front of the wave inflates, the cell at the back of the wave starts to deflate, so at any one time only a small area is being compressed.

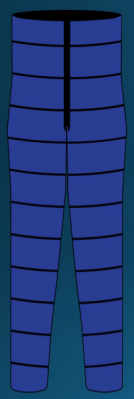
PNEUMATIC COMPRESSION SETTINGS

PERISTALTIC PROGRAMMING:

- PERISTALTIC MODE MAY BE BETTER TOLERATED FOR PATIENTS WITH HAND/FOOT NEUROPATHY BECAUSE PRESSURE IS NOT SUSTAINED DISTALLY
- PERISTALTIC MODE AT HIGHER PRESSURES MAY BE BEST CHOICE FOR TREATING PROXIMAL FIBROSIS (AXILLA, BREAST, TORSO)

Pre-Therapy

Pre-therapy can be added to the beginning of the Sequential or Peristaltic cycles. Pre-therapy treats the proximal areas first, decongesting them so that they can receive the lymph fluid that is mobilized during the Sequential or Peristaltic cycles. A specific pattern is applied, and the duration will vary according to garment size, lasting between 5-12 minutes.



Optimal Pre-therapy

Pressure 40 distally in gradient of 1 mmHg.

Inflation cycle by chamber:

11-12

11-12

10-11-12

10-11-12

9-10-11-12

9-10-11-12

8-9-10-11-12

8-9-10-11-12

It will continue this pattern until you get to two cycles of

1-2-3-4-5-6-7-8-9-10-11-12

Post-Therapy:

Exclusive to PCD-52

Post-therapy can be added to the end of the Sequential or Peristaltic cycles. Post-therapy provides a "focused" treatment for either the proximal, medial or distal areas, according to user selection. It then follows it with a wave-like "sweep" over the entire treated area, according to the principles of manual lymph drainage. Post-therapy duration is 5 minutes, at the zone pressure that has been elected for the Sequential or Peristaltic cycles.

CONTRAINDICATIONS FOR PUMP USE:

- KNOWN/SUSPECTED DVT OR PULMONARY EMBOLISM
- INFLAMMATORY PHLEBITIS
- ACUTE INFECTION OF THE AFFECTED LIMB
- DECOMPENSATED CARDIAC FAILURE
- SEVERE ARTERIOSCLEROSIS, ISCHEMIC VASCULAR DISEASE
- ANY CIRCUMSTANCE WHERE INCREASED VENOUS AND LYMPHATIC RETURN IS UNDESIRABLE
- DON'T TREAT ABDOMINAL AREA DURING PREGNANCY

CAUTION REQUIRED:

- PERIPHERAL NEUROPATHY
- PAIN OR PARESTHESIA
- OPEN WOUNDS
- FRAGILE SKIN, SKIN GRAFTS
- EXTREME LIMB DEFORMITY
- COLOSTOMIES, MEDIPORTS OR OTHER APPLIANCES
- USE LOWER PRESSURES WITH PVD
- MONITOR PATIENTS WITH HEART DISEASE

COMPRESSION

- BANDAGING
- WRAPS
- ELASTIC COMPRESSION
- QUILTED COMPRESSION
- BULK IS BETTER FOR EDEMA CONTROL AND FIBROSIS:
  - MORE IS MORE: "BOOT CAMP" PERIOD

MODALITIES

- LASER
- ELASTIC TAPING
- THERMAL MODALITIES
- VIBRATION (HAND HELD, PLATFORM)
- NEGATIVE PRESSURE
- DRY NEEDLING
- HIVAMAT