



Dr. Herbst - New Patient Visit

Patient Name: Allen , Nina

DOB: 1969-10-24

Age: 51

Referring Physician: Andrew Tak

Phone number: (916) 646-4583

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Joel Crawford, MD, RPVI; Vascular Surgery

Phone: 916-773-8750

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Pharmacy Name: Walmart

Pharmacy Number: 916-534-1157

Pharmacy Address: 6005 Madison Ave, Carmichael, CA 95608, United States

CC: Lipedema. Confirm a diagnosis, get recommendations for medical and surgical treatment

HPI: Nina Allen is a 51 year young female with a history of Lipedema, a loose connective tissue disease. Her Lipedema was diagnosed in June 2021 by her vascular doctor, Dr Crawford. Her Lipedema began in February 1994. Elevating legs, chiropractic adjustments, stretching, limiting driving, and sleep helps her Lipedema. Driving more than 30 minutes, sitting for extended periods of time, walking, strenuous exercise, typing, and standing makes her Lipedema worse. Her Lipedema is constanly progressing. She has never had a large butt and now she does. She feels like she is in system malfunction.

She also has lower abdomen swelling.

Onset: During her pregnancy with her daughter in 1994. She felt her body fill with fluid and she gained weight. After her daughter she was not able to lose the weight for about two years. She was diagnosed with a goiter by an Endocrinologist and she went on medication and finally got off the medication as her TSH levels normalized. Her symptoms persisted such as dry skin, brittle hair, swelling. Before she was pregnant she weighed 124 lbs and her body fat was 17%. The shape of her body changed when she was pregnant. She was never a big eater and never had weight issues before her pregnancy. Now it hurts to walk with joint pain and lower back pain. She now has pain that she never experienced. If she does vigorous exercise she suffers for weeks and it never ceases. She is on Motrin for weeks after exercising.

Swelling? Yes she was told she had edema and was given compression stockings/socks and it felt like it was locking in the swelling and not reducing it. For years they told her she had edema and to reduce salt. She does not eat red meat, drink or smoke and she had not changed her eating habits. Her varicose veins worsened. She does not have venous insufficiency by venous duplex ultrasound supine and standing. Her veins are definitely painful and her legs feel tired.

Swelling worse during summer? Heat bothers her and slows her down and she feels a fatigue.

Swelling worse when standing? Yes

Swelling worse when sitting? Yes

Limbs tight and heavy especially at end of day? Yes tighter and heavier.

Do you elevate your legs? Yes it helps but they have to be elevated or else they will swell. It feels like fluid. If she does a lot of leg exercises she will fall asleep as she feels she does not have enough oxygen.

Does swelling resolve with elevation or sleeping overnight? Yes a little better. When she sleeps she wakes up stiff.

Areas with lipedema are unaffected by caloric restriction? Hips, buttocks, thighs and her arms. Her arms used to be muscular but now she cannot build muscle and the shape of the arms have changed.

Reduced ability to get around (ambulation)? Yes especially if she drives for work. For days she is stiff and feels like she needs oil to lubricate her joints and she feels knock knees because everything is tight and constricted.

Any areas of your body that are colder than other parts? Feet and hands.

Any decrease in social activity? Yes. If she is in a lot of pain she will not go anywhere. She may also be too tired. She expends a lot of energy working and doing basic stuff. She does not have energy for more.

Diet: Ketogenic diet, Plant-Based, Whole Food, green smoothie diet. First meal 12PM, last meal 9:30PM. 2 meals, 2 snacks.

Exercise: Low impact cardio, 25-30mins, with sweating. Just got a whole body vibration machine and it helped but she is easing into using it. She feels when she is exercising the swelling should move but she feels the fluid is trapped with no way to release it. The more she does exercise the more this becomes an issue. She has not tried the pool yet.

Pain

Average Daily Pain Score (1-10): 9

Worst Daily Pain Score (1-10): 11

Lowest Daily Pain Score (1-10): 6

Pain is in the: Calves, knees, thighs, shoulders, elbows, hands, buttocks, lower back, stomach, feet

Conservative Therapy

Compression Garments: Compression socks: periodically (when flying); no improvement

Sequential Pneumatic Compression Pump: None

Manual Lymphatic Drainage Therapy: October 2019 - Lymphatic drainage massage: discomfort (during and after), severe headache for 5 days. During the massage the therapist could pinpoint her areas of pain.

Deep Tissue Therapy: None

Weight

Any history of weight gain: Yes, 1994: weight gain following birth of 2nd child continued progressing with small weight loss which quickly reached plateaus regardless of diet and exercise

Any history of weight loss: Yes, stomach and arms mainly; these areas area also very difficult to lose weight from

Ever use of the following meds

Phentermine: No

Dextroamphetamine: No

Adderall: No

Metformin: No

MEDICAL HISTORY

First Menses: 11

Menopause: 48

3 Pregnancy(ies)

2 Live Births

1995: Goiter

1998: Deviated Septum

2020: Gallbladder Removal

Chronic: Anemia

2011: Pneumonia

Hypothyroid

Migraine headaches

Polycystic Ovarian Disease - she is unsure of this diagnosis. But after her thyroid issues she had a heavy menses and felt like she was hemorrhaging at times. She feels there is hair on her chest. She has had ultrasound of her ovaries and they are fine.

Varicose Veins

SURGICAL HISTORY

- 1978: Eye surgery - 1990: C-section - 1998: Deviated Septum Surgery - 2020: Gallbladder Removal
Emergency Caesarean section in August 1990 Complications from fetal stress

MEDICATIONS

Allergies: - Erythromycin - Vicodin/Hydrocodone

Medications: N/A

Supplements:

Vitamin C / 500 MG / 2x Day

Vitamin D / 2,000 IU / 1x Day

Vitamin B12 Complex / 1,017 MG / 1x Day

Multivitamin / 1x Day

Iron / 1000 MG / As tolerated (1x Day max)

[Medications were reviewed]

SOCIAL HISTORY

Smoking: N/A

Alcohol: 0

Any other drugs: N/A

FAMILY HISTORY

- Diabetes - Cancer - Multiple sclerosis - Cushing's syndrome - Hypertension - High Blood Pressure - B12 Deficiency - Vitamin D Deficiency - Anemia

Her paternal grandmother likely had lipedema. She had the typical body shape but she said she was thin in the past.

Sister had Cushing's disease s/p surgery.

REVIEW OF SYSTEMS

General: Weight gain, weight loss, difficulty sleeping. No complaints of: flu-like symptoms

HEENT: Difficulty swallowing, neck feels swollen, dry eyes, dry mouth. No complaints of: thick skull fat

CV: Chest pain. No complaints of: palpitations

Dermatology: Easy bruising, itching in skin/tissue, water trickling under skin, burning sensations, keloids, stretch marks.

Endocrine: Fatigue 9/10, cold feet/hands, pre-diabetes. No complaints of: feeling thirsty all the time

Gastrointestinal: Bloating, stomach/intestinal pain, early satiety. No complaints of: diarrhea, constipation, nausea, vomiting

Genitourinary: Incontinence, Nocturia 3 times. No complaints of: pain with intercourse

Immunology/Infectious Disease/Allergy: Inflammation in blood, Allergies (Erythromycin, Vicodin/Hydrocodone)

Musculoskeletal: Muscle aches, muscle weakness, tight tendons, muscle cramps, joint aches, low back pain, flexible joints

Neurology: Vibrations in tissue, vertigo (once in 1997), hearing loss, poor concentration, numbness (hands, feet, legs)

Pulmonary: Sleep apnea not on CPAP - it is getting better. She used to wake up at night coughing but that calmed down. She was started on high dose magnesium which helped her muscles relax. She rarely snores now. No complaints of: frequent congestion, shortness of breath

Psychiatry: Depression, anxiety, sexual/physical/emotional trauma

Vascular: Water retention, swelling, dark skin on lower legs. No complaints of: blood clot

Other symptoms or concerns: See HPI

PHYSICAL EXAM

BP: NA/NA HR: 52 Weight: 226 Height: 5'1" BMI: 42.79 TEMP: 97.5

Waist (cm): 97.5

Hips (cm): 147

Waist-to-hip-ratio:

A WHR of ≥ 0.85 cm is suggestive of obesity in women (World Health Organization, 2011). A value of

General: Woman in no apparent distress

Gait: Legs rub together from groin to knees

HEENT: PERRLA; EOMI; does not wear glasses

Neck: No thyroid enlargement or nodules

Heart: Regular rate and rhythm; no murmurs, rubs or gallops

Lungs: Clear to auscultation

Abdomen: Non-distended, soft

Vascular: Stemmer negative on the hands and feet; No pitting edema; no evidence of acrocyanosis

LOOSE CONNECTIVE (FAT) TISSUE EXAM

Head and Neck

Cranial fat: Normal

Neck: Acanthosis nigricans: Yes

Lymph nodes: Normal

Supraclavicular fat: Increased

Back

Dorsocervical fat pad: No

Folds of connective tissue on the sides of the back or under the bra: Yes two folds both with nodular tissue and fibrotic tissue inside

Lordosis: Yes

Shelf of tissue above the buttocks: Yes that is tender; buttocks are also tender deep

Chest

Increased long dark hairs anterior chest

Arms

Axillary: Acanthosis nigricans: Yes

Axillary: Full and tender: Yes

Increased tissue upper arm: Yes

Palpable nodules upper arm: Yes

Increased tissue lower arm: Yes

Palpable nodules lower arm: Yes

Wrist cuff: Yes right and left side

Hand fat base thumb: Yes

Hand fat between MCPs: Yes

Stemmer hand: Negative

Heavy upper arms: No

Abdomen

Palpable nodules: Yes

Panniculus: Grade 3: the panniculus reaches down to the upper thigh

Heavy panniculus: Yes

Palpable nodules suprapubic: Yes

Legs

Striae: Yes

Mattress pattern thigh tissue: Yes

Palpable nodules thigh tissue: Yes

Fat overhanging knee: Yes

Fat covers knee: Yes
Fat covers shin: Yes some of the shin
Medial knee lobule: Yes
Stovepipe legs: No
Increased tissue lower leg: Yes
Palpable nodules calves: Yes
Ankle cuff: Yes
Fat around lateral malleoli: Yes
Fat around medial malleoli: Yes
Fat around Achilles: Yes
Fat on top of foot: Yes
Stemmer foot: Positive
Piezogenic papules: No
Flat feet: Yes

Vascular Exam

Telangiectasia/Spider Veins: Yes
Visible Varicose Veins: Yes by thermography on the calves and extending to the outer thighs and into the abdomen
Non-pitting edema: Yes
Corona phlebectatica: No
Pitting edema: Yes

Areas of Hypothermia

Arms: Yes
Legs: Yes calves
Buttocks: No
Hips: No

Joints

Valgus of knees: No
Varus of ankles: No

General

Tissue tender in areas affected: Yes
Hands and Feet Not Affected: Hands not affected but lymphedema present after traveling; feet are relatively less affected than the legs
Bruising currently: Right arm
Fibrotic Tissue: Yes in the nodules and in the folds of the tissue on the back and the panniculus
Heavy Tissue: Yes buttocks, hips, thighs, breasts

Beighton Score:

5th digits - 0/2
Thumbs - 0/2
Elbows - 2/2
Knees - 2/2
Hips - able to bend and touch the floor keeping the legs together and straight = 1
Score: = 5/9

Diagnostic Criteria for lipedema

Female: **Yes**
Bilateral and symmetrical manifestation with minimal involvement of the feet: **Yes**
Minimal pitting edema: **Yes**
Negative Kaposi–Stemmer sign: **Yes of the hands but not of the feet today**
Pain, tenderness on pressure: **Yes**
Easy bruising: **Yes**
Persistent enlargement after elevation of the extremities or weight loss: **Yes**
Arms are affected 80% of the time: **Yes**

Hypothermia of the skin: **Yes**

Swelling worsens with orthostasis in summer: **Yes**

Lipedema tissue unaffected by caloric restriction, exercise, bariatric surgery: **Yes**

Vascular manifestation such as cherry angiomas, telangiectasia, venous disease: **Yes**

Does the patient meet criteria for lipedema? **Yes**

Labs:

NA

The lower extremity functional scale (**LEFS**) is a measure of disability for the legs. Lower scores indicate more dysfunction.

Score = 15/80

Five Questions for Hypermobility: 1/5

A positive answer for two or more questions has a sensitivity of 91%, a specificity of 75% for predicting hypermobile joints (BMC Musculoskelet Disord. 2020; 21: 174).

ASSESSMENT

1. Lipedema Stage 3 Type III and IV

Lipedema of the abdomen

Lipolymphedema

Lipedema is a congenital enlargement (hyperplasia of the adipose tissue) of the loose connective (fat) tissue on the legs almost exclusively seen in women by the third decade. According to an epidemiologic study by Földi E and Földi M, lipedema affects 11% of the female population. Lipedema was initially described by Allen and Hines in 1940; its etiology remains unknown and it remains under-diagnosed. Classically women with lipedema have disproportionate bodies with larger legs and hips than arms and waist. In 1951 Wold et al. analyzed 119 cases and provided the diagnostic criteria for lipedema:

- 1) Almost exclusive occurrence in women
- 2) Bilateral and symmetrical manifestation with minimal involvement of the feet
- 3) Minimal pitting edema; the Kaposi-Stemmer sign is negative
- 4) Pain, tenderness on pressure
- 5) Increased vascular fragility; easy bruising
- 6) Persistent enlargement after elevation of the extremities or weight loss
- 7) Arms are affected 80% of the time
- 8) Hypothermia of the skin
- 9) Swelling worsens with orthostasis in summer
- 10) Unaffected by caloric restriction

The stage of disease refers to how the skin and tissue appear visually:

When the skin is still smooth, the lipedema is stage 1.

When the skin and tissue have indentations in a mattress pattern, the lipedema is stage 2. Lipedema stage 3 has larger out-pockets of tissue.

The types of lipedema refer to the location of the fat:

Type I: In the area of the buttocks and hips (saddle bag phenomenon)

Type II: Buttocks to knees, with formation of folds of fat around the inner side of the knee

Type III: Buttocks to ankles

Type IV: Arms

Type V: Legs

In lipedema, there are increased macrophages in tissue, a microangiopathy (leading to increased bruising), dilation of subdermal capillaries which can be seen as telangiectasias and petechiae on the skin, dilation and leakage of lymphatic vessels in the subcutaneous fat - leaking lymphatics into subcutaneous fat increases growth of adipose tissue in mouse models.

Diuretics such as Lasix concentrate proteins in the interstitium increasing the work load of the lymphatic system.

Do not use diuretics.

Corticosteroids should be avoided as they weaken blood vessels (and lymphatics) and cause a rebound increase in adipose growth once stopped.

For any surgery, there must be professional manual lymphatic drainage at minimum one week before and for four

weeks after the surgery - longer if there is a slow recovery. In lipedema and lymphedema (lymphatic dysfunction), there is difficulty in handling all the fluid and inflammation after surgery. This means there is a need for hands on MLD from a trained practitioner. Mismanagement of MLD after surgery would risk the development of difficult to control lymphedema. Adequate MLD after surgery is standard of care (<https://pubmed.ncbi.nlm.nih.gov/34049453/>).

2. Varicose veins.

3. Hypermobile Joint Syndrome: Generalized joint hypermobility (GJH) or Ehlers Danlos Syndrome hypermobile type (EDS-HT), better known as hypermobility spectrum disorders, are conditions causing joint laxity. The gene for either condition is not known although tenascin-X made from the TNXB gene has been found in some families. Tenascin-X plays an important role in organizing and maintaining the structure of tissues that support muscles, joints, organs, and skin (connective tissues). In particular, studies suggest that tenascin-X helps regulate production and assembly of certain types of collagen. Collagens are a family of proteins that strengthen and support connective tissues throughout the body. Tenascin-X is also involved in regulating the structure and stability of elastic fibers, which provide flexibility and stretchiness (elasticity) to connective tissues.

Fat tissue is known as loose connective tissue which consists of sheets of connective tissue called fascia, fibers including collagen and elastin, fat cells, immune cells, fibroblasts, blood and lymph vessels. Between cells water is bound to glycosaminoglycans (sugar molecules). Blood vessels, nerves and lymphatics pass through fat on fascia highways and fat lobules slide on thin wet fascia ropes between skin and muscle accommodating movement.

When there are changes in the genes causing connective tissue proteins to be differently formed, skin loses its ability to maintain its shape, blood vessels leak, lymph vessels dilate and fail to pump, and the fascia ropes tighten and inhibit movement. Fluid, protein, and cell waste sit in fat tissue, providing all the nutrients and growth factors to make fat cells grow and proliferate in this nutrient-rich environment. Joints, muscles, tendons, and ligaments become looser and more fragile.

Not everyone with hypermobile joints develops symptoms. Different genetic changes may also result in similar symptoms. Fat disorders, including lipomas, may result from changes in genes important in mobility.

The diagnosis of EDS-HT based on history and a clinical exam. Women with hypermobile joints have a risk of osteoporosis and should have a DEXA scan for bone disease at menopause. An echocardiogram should also be performed to determine if the aortic root is dilated. There are also a lot of gastrointestinal issues in EDS.

Find out more here: <https://www.ehlers-danlos.com/2017-eds-classification-non-experts/gastrointestinal-involvement-ehlers-danlos-syndromes/>

You can read more about EDS-HT here: <http://www.ncbi.nlm.nih.gov/books/NBK1279/> and here: <https://www.cda-adc.ca/jcda/vol-67/issue-6/330.html>

What are some findings in GJH or EDS-HT?

1. Celiac Disease: There is an increased prevalence of potential celiac disease. Serological screening of celiac disease is recommended for to rule out organic problems in the presence gastrointestinal symptoms in patients with GJH or EDS-HT.¹

2. ADHD: GJH may represent a marker for an underlying systemic disorder involving both connective tissue and the central nervous system. Specifically, attention deficit hyperactivity disorder (ADHD) was significantly associated with GJH.²

3. Complex Syndromes: EDS has been linked to Chiari Malformation, Postural Orthostatic Tachycardia Syndrome, and Mast Cell Activation Syndrome.³

4. Ocular complications: Find more here: totaleyecare.com

4. Possible congenital adrenal hyperplasia (CAH) adult onset.

PLAN

1. **Manual therapy:** Find a certified lymphedema therapist (CLT) who can provide manual lymphatic drainage therapy to reduce fluid, deeper manual or instrument assisted soft tissue therapy to reduce fibrosis, educate on skin care and compression and discuss the use of a sequential pneumatic compression pump.

<https://klosetraining.com/therapist-directory/>

<https://lymphnet.org/find-treatment>

<https://www.clt-lana.org/>

2. **Compression garments:** Leggings high waist down to the foot. I suggest lower level of compression leggings CZ Salus, Bioflect or Solidea and you can layer with your lower leg stockings. Order provided.

3. **Lymph Pumps (E0652):** I recommend two pumps for treatment of rare adipose disorders: Lymphapress Optimal (877-316-8458) or Flexitouch (866-435-3948). The benefit of the Lympha Press is you can treat both legs

at one time. These are the only two pumps I allow my patients to use. It is not standard practice to prescribe sequential compression pumps (SCDs) usually reserved for prevention of deep venous thromboses or for treatment of cardiovascular edema (E0650; E0651). In the latter two situations, the lymphatic vessels are intact and pump normally. As a consequence of using SCDs in RADs, SCD pumps push the fluid up the leg into the abdominal area where it accumulates due to lymphatic dysfunction. As this fluid sits in the tissue with all its nutrients and protein, fat grows. From published data, we know that lymph makes fat grow (Nat Genet. 2005 Oct;37(10):1023-4). With the Lymphapress or Flexitouch, the abdomen is treated along with the leg and the chest is treated along with the arm preventing dangerous pooling of lymph fluid. A E0652 device with a segmented, multi-ported pump allows for individual pressure calibration at each port. This allows the patient to alter pressure in areas of severe pain as found in Dercum's disease or lipedema while obtaining a compression sleeve that treats the abdomen and/or chest. I will order you a Flexitouch pump for the arms and legs.

4. Nuclear medicine lymphangioscintigraphy exam ordered.

5. **Labs:** Check 17-OH-progesterone, insulin and glucose and AM Cortisol and testosterone and androstenedione.

6. Agree with lipedema reduction surgery to treat the lower body, arms and abdomen. Consider Dr. med Manuel Cornely:

Address: Kaiserswerther Str. 296, 40474 Düsseldorf, Germany

Hours: **Closed** · Opens 8AM Mon

Phone: [+49 211 352825](tel:+49211352825)

Appointments: samedi.de

ICD-10 codes for this visit

R60.9 Lipedema

I89.0 Lymphedema

I86.8 Varicose veins

M79.605 Pain in the left leg

M79.604 Pain in the right leg

M79.601 Pain in the right arm

M79.602 Pain in the left arm

Q79.62 Hypermobility Ehlers Danlos Syndrome

G89.4 Chronic pain syndrome

R10.9 Unspecified abdominal pain

This visit was 60 minutes with >50% time spent counseling on lipedema and other causes of fat tissue growth and possible treatments that may help Nina.

Electronically signed by Karen L. Herbst, MD, PhD 2021-08-13 8:12 AM

Karen L. Herbst, MD, PC

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