

## CASE STUDY

# The Mystery is Solved: A Case Study of Delayed Diagnosis and Treatment

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Laura (pseudonym for the patient) is a 48-year-old woman referred to our clinic, presenting with a first-time diagnosis of (complicated) primary lymphedema of the right lower extremity. She shared her story, reporting that she couldn't remember a time when her right foot and ankle were not noticeably swollen. Laura remembers doctor visits and "special shoes" that her parents would have to "save up to purchase." Although Laura struggled to keep up with the other kids and walked with a limp, no treatment interventions were offered until Laura turned 12 years old. In 1980, she presented with a painful, palpable lump in the right inguinal region. The doctors reassured Laura's parents that she did not have cancer, as they had feared, but rather a "rare, mysterious condition" that caused problematic swelling and an "infected" lymph node. She was treated for the infection and was directed to begin wearing compression stockings. As she grew into adulthood, it was still a challenge to find shoes that fit. The compression stocking and ill-fitting shoes were uncomfortable, especially during the long hours of nursing school. To make matters worse, Laura was hit by a drunk driver on the day after she passed her state nursing board exams. The accident caused extensive injuries, including multiple right leg fractures. She spent the next nine months wheelchair-bound, followed by six more months ambulating on crutches. She tried to resume her nursing career, but was challenged with pain, reoccurring episodes of cellulitis, and increasing

right lower leg swelling over the years. Her primary physicians offered limited solutions to address these issues. She is now married and helps with the family business. She is also an active mother of three children, one of whom is disabled.

Laura shared that she felt that the "rare, mysterious condition" of her right foot, rather than the accident, was the main cause of an unresolvable cycle of disability. In July of this year, an unexpected cardiac condition resulted in a consultation with a cardiovascular specialist. The cardiologist noted the presentation of Laura's right foot and lower leg.



He ordered additional tests, including a lymphoscintigraphy test. This test was "an imaging technology using contrast material to detect lymphatic malfunction."<sup>1</sup> In Laura's case, the test documented the imaging of lymphatic abnormality and dysfunction of the right foot, with delayed response. It was concluded that the abnormality was distal to the orthopedic injuries sustained in the accident, determining that Laura's mystery condition was congenital primary lymphedema of the right lower leg<sup>1</sup>, complicated by trauma. The mystery of the swollen foot was solved. Laura was relieved to learn that primary lymphedema, although incurable, can be treated and managed. She was referred to our lymphedema treatment clinic to begin occupational therapy. Her hope was to "fit into a normal size pair of jeans and a good pair of shoes."

Upon arrival for the initial occupational therapy evaluation, Laura was accompanied by her husband. She complained of metatarsalgia, ankle stiffness, and pain to the right knee during ambulation. Her foot pain was centered over the metatarsal region. Her toes did not engage the floor during weight bearing and ambulation due to significant swelling to the sole of the foot. There was limited flexion and extension of the toes due to a dorsal foot lobule and increased circumferential edema girth. Characteristic of progressive stage II lymphedema, the dorsal foot resisted indentation with palpation. The retro-malleolar region was densely indurated.<sup>2</sup> She offered a positive Stemmer sign of thickening, immobile skin folds to the right toes.<sup>2</sup> There was also a significant circumferential girth differential in right-to-left bilateral comparison of the entire lower leg (Figure 1).



Laura's right ankle seemed unstable as she walked and appeared to collapse with inversion. She complained that walking up the incline ramp or moving from sitting to standing was difficult and painful to the right knee. She limped due to the pain and muscle imbalance. When observed, Laura assumed a moderate hyperextended knee pattern and stood with a "locked knee" position, suggesting weakness of the gastrocnemius and quadriceps muscle group.<sup>4</sup> This may have been a residual effect of the orthopedic injuries sustained during the motor

vehicle accident as a young adult and was complicated by the lymphedema.

Laura and her husband were educated in the principles of complete decongestive therapies (CDT) and introduced to the components of manual lymph drainage (MLD)<sup>3</sup>, therapeutic compression, and exercise to support lymphatic dynamics to promote lymphostatic decongestion. Skin care needs were also addressed and introduced as part of her developing plan of care. Although a plan of treatment consisting of participation in therapy three to five times per week seemed ideal, Laura's need to care for her disabled child created a challenge in time management. It was mutually agreed that she would participate in direct treatment sessions two times a week, but would complete structured, self-directed MLD every day as she and her husband showed competency of the treatment method. They received their first MLD instruction through demonstration and handouts that offered step-by-step diagrams designed for Laura's MLD program. Laura's husband also videotaped the demonstration for a visual aid at home.



The MLD treatment followed traditional sequence designs<sup>3,5</sup>, including anatomical pre-treatment of the neck, viscera, and deep abdomen, as well as the right axillary nodes. Establishing the right inguino-axillary anastomoses and beginning treatment of the left inguinal lymph nodes was the next step. This was followed by anterior inter-inguinal anastomoses. Areas of density were given special attention with deeper techniques to soft the tissues, promoting lymphostatic decongestion. The treatment extended the entire length of the right leg and included the posterior aspect.

In partnership, Laura and her husband learned how to apply compression in the first phase of treatment. This included a chip bag-like foam over the dorsal right foot and the ankle, where fibrosis restrictions were problematic. A short-stretch, layered compression bandage system was employed.<sup>3,5</sup> Laura also continued using principles of skin care to keep her skin moisturized.

To address Laura's complaint of right leg weakness and to reduce her discomfort while exercising or walking, she was instructed in a home program that included the application of elastic taping to enhance the strength of the gastrocnemius and quadriceps. Although there is ongoing discussion regarding its role in lymphedema reduction, elastic taping is recognized for its therapeutic value in the treatment of musculoskeletal pain and weakness.<sup>6-7</sup> Incorporating elastic taping offered Laura a way to enhance muscle strength, support lymph drainage, and reduce pain simultaneously.

After four weeks, Laura is encouraged by the improvements of lymphedema reduction to her right leg. Her leg pain is gone and she is walking without a limp. Laura has reached her first goal of fitting into "skinnier jeans." Although she continues participating in treatment, she was recently fitted with a flat-knit compression stocking and a custom compression piece for the toes to use intermittently for an occasional "break" from the bandages, although she will still wrap every night. She is focused on fitting into those shoes!

**Discussion:** Primary lymphedema is an underdiagnosed, debilitating condition that can directly or indirectly impact patient care. The uncertainty as to when to make the diagnosis of lymphedema, what treatment approaches are most effective, and the disconnect around recognizing this condition's potential influences on progressive disability continues to propagate the idea that it is a "rare, mysterious condition" with few treatment options beyond uncomfortable compression garments. We must continue to support every effort to educate the medical community regarding the advances in research, technology, and developing treatment interventions that are now available to "de-mystify" lymphedema so that those who struggle with this condition are offered a chance for an improved quality of life.

Figure 1

<b>Anatomical Landmark</b>	<b>Right Initial</b>	<b>At week four of treatment</b>	<b>Left</b>
Dorsal Foot	30.7 cm	26.7 cm	24.0 cm
Retro-Malleolar/ heel	39.2 cm	35.2 cm	31.5 cm
Ankle	29.0 cm	26.0 cm	22.0 cm
6" proximal to the ankle	40.0 cm	36.8 cm	32.0 cm
10" proximal to the ankle	48.6 cm	42.8 cm	39.0 cm
Knee Crease	42.0 cm	38.7 cm	35.0 cm
9" proximal to the knee	52.0 cm	50.8 cm	49.0 cm

## References

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