Let's talk genital lymphedema

Improving ways to diagnose and treat the taboo area

***Pathways* recently had an opportunity to chat with Shelley DiCecco regarding a much-needed area of lymphedema that has not been written about very much. She has been kind enough to provide our readers with answers to typical questions and included some practical charts and tools to help health professionals address genital lymphedema.**

Why is genital lymphedema not given much coverage?

In many countries we are taught from an early age that our genitals are not to be discussed or shown to others, that the topics of genitals and sex are considered taboo. Even as health professionals we are often advised to only go there if absolutely necessary. Unfortunately, these ingrained notions carry over into the evaluation and treatment of lymphedema. Accurate statistics on the percentage of individuals living with genital lymphedema are lacking. There are several reasons for this: 1) lymphedema in general is under diagnosed; 2) patients and medical staff feel uncomfortable when discussing the genital region, and 3) there is a widespread lack of knowledge on how to properly assess and treat genital lymphedema. This article intends to improve communication between patients and healthcare professionals (HCP), by improving the comfort level of HCPs in addressing genital lymphedema.

Can genital lymphedema be primary or secondary?

Genital lymphedema is associated with both primary and secondary lymphedema. In primary lymphedema the genital involvement typically accompanies edema in other areas of the body, particularly in one or both lower extremities. Males are reported to have a higher incidence of primary genital lymphedema than females.[1,2] A study of 138 children with lymphedema from Children's Hospital Boston found that males were seven times more likely to have genital lymphedema as compared to females.[2] Is this accurate or is it just that female genital lymphedema, especially in children, is more difficult to diagnose? The study by Schook et al also noted that 12% of the children with initial lymphedema isolated to the leg(s) later developed genital lymphedema.[2] Secondary genital lymphedema can result from filariasis, cancer treatment, lymphedema in adjacent areas, obesity, trauma, or surgeries on the lower trunk and/or genitals. Some less common causes can include pregnancy, Crohn's disease, long distance cycling, hormonal therapies, hydradenitis suppurativa, STDs, and other co-morbidities.[3-6]

What are the characteristics of genital edema?

Genital lymphedema in males can involve portions of the penis, scrotum, or both. It is not uncommon for the penis to become buried or "engulfed within the prepubic skin" with significant scrotal edema.[6] In females, the involvement includes the labias, the vulvar vestibule, clitoris, and/or the perineal tissue. A less evaluated area for females is the vagina. Inflammation can evolve from the vagina down into the labia(s); early detection/treatment may prevent the progression. The edema may only involve one side of the genital region in females, for example only one labium. For both sexes, other structures frequently involved include the mons pubis, anus region, inner thighs, gluteal region/buttocks, and lower abdomen.[5, 7-9]

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What are the symptoms associated with genital lymphedema?

Symptoms common with genital lymphedema for both sexes include urinary dysfunctions, bowel dysfunction, pain in the genital region, skin breakdown, wounds, lymphorrhea, odor, disfigurement, sexual dysfunction, frequent bouts of cellulitis, associated difficulties completing daily hygiene activities, and decreased quality of life.[1,4,7-11] A study by

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> Obese individuals with BMIs higher than 40 have an increased incidence of genital lymphedema.

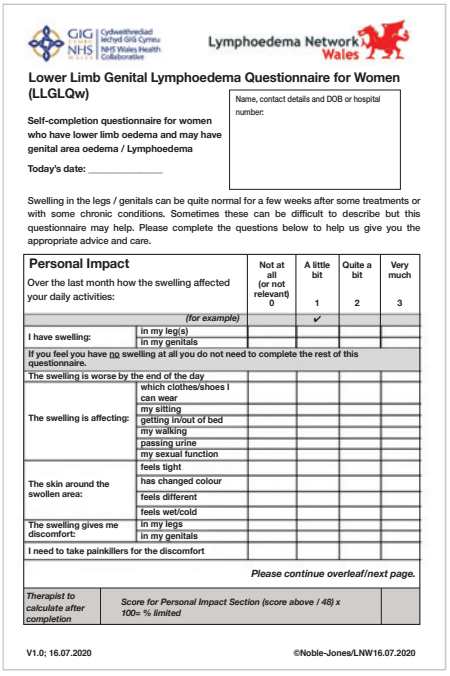
Schook et al involving 25 males with genital lymphedema noted 60% reported some form of genitourinary symptoms.[8] Psychological distress is common with lymphedema, and especially with genital lymphedema. The distorted shape, the impact on intimacy with others (inability to participate, reduced libido, embarrassment, and/or fear on the part of the partner), the fear of odor and/or leaking, and the noticeable alteration in gait due to the size of the genitals all add to psychosocial stress.[1,4,7,8,11] All symptoms need to be acknowledged and addressed when possible, by HCPs.

What is the connection of adipose tissue and genital lymphedema?

Obese individuals with BMIs higher than 40 have an increased incidence of genital lymphedema. This can be caused by pathophysiological damage to the lymphatic system from the adipose tissue and/or from the weight of the pannus abdomen on the inguinal nodes.[12] All adipose tissue releases adipocytokines which can increase inflammation and interfere with lymphangiogenesis or cause lymphatic vessels to become leaky.[13,14] Chronic inflammation leads to the formation of additional adipose tissue via adipocyte stimulation. Studies on the relationship between lymphedema and adipose tissue demonstrated increased levels of adipogenic transcription factors or adipocytokines.[12,15,17,19]

Lymphedema in the lower abdomen and/or genital region near the mons pubis may increase the deposition of adipose tissue at a higher rate than in other regions due to the proximity to the protective adipose tissue at the pubic bone or mons pubis area. Once abnormal adipose tissue from inflammation is present, it can only be removed via surgical intervention. This is one of the reasons why it is important to address trunk and/or genital lymphedema early and not to avoid this area due to HCP discomfort with treating this area.

IMAGE 1



Lower Limb Genital Lymphoedema Questionnaire for Women (LLGLQw)

Name, contact details and DOB or hospital number:

Self-completion questionnaire for women who have lower limb oedema and may have genital area oedema / Lymphoedema

Today's date: _____

Swelling in the legs / genitals can be quite normal for a few weeks after some treatments or with some chronic conditions. Sometimes these can be difficult to describe but this questionnaire may help. Please complete the questions below to help us give you the appropriate advice and care.

Personal Impact Over the last month how the swelling affected your daily activities:		Not at all (or not relevant) 0	A little bit 1	Quite a bit 2	Very much 3
(for example)			✔		
I have swelling:	in my leg(s)				
	in my genitals				
If you feel you have no swelling at all you do not need to complete the rest of this questionnaire.					
The swelling is worse by the end of the day					
The swelling is affecting:	which clothes/shoes I can wear				
	my sitting				
	getting in/out of bed				
	my walking				
	passing urine				
	my sexual function				
The skin around the swollen area:	feels tight				
	has changed colour				
	feels different				
	feels wet/cold				
The swelling gives me discomfort:	in my legs				
	in my genitals				
I need to take painkillers for the discomfort					

Please continue overleaf/next page.

Therapist to calculate after completion	*Score for Personal Impact Section (score above / 48) x 100= % limited*	

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Front page of the genital lymphedema tools for females.

What are the elements of a comprehensive evaluation?

Subjective:

A main limitation in the evaluation of genital lymphedema is the discomfort level of the HCP and the patient in discussing this region. A study by Nobel-Jones et al noted both HCPs and patients preferred if the other brought up genital involvement during the visit. Both agreed this hesitation could lead to a significant delay in treatment of the genitals.[5,20] A screening tool for the patient to complete prior to seeing the HCP addressing all the possible symptoms can facilitate the initiation of the conversation. The answers to the tool still need to be discussed in detail with the patient. For even with a tool, patients still may not answer correctly or honestly, or they may not understand the questions. Some feel incontinence means complete loss of urine, not just involuntary loss of one or more drops of urine. The study by Noble-Jones et al noted that patients felt the use of a screening tool could provide the impression that the patient is not alone in the symptoms.[20] HCPs should be asking clarifying questions regarding symptoms with all medical conditions, especially genital lymphedema, to include: when do the symptoms occur, where exactly do they occur, what makes them worse, and what makes them better? For sexual dysfunction, the answers to the clarifying questions can provide important information on the exact nature of involvement, which can help guide the objective portion of the assessment. For example, in females, pain with initial penetration during intercourse involves the external genitals (vulva vestibule or labia) and pain with deeper activity involves more of the vaginal tissues. Noble-Jones et al developed screening tools for both males and females to address these symptoms during evaluation and to provide a means of initiating discussion into this difficult conversation.[5,20] The full tools can be downloaded for free at **www.lymphed.com/edematool**.

Objective:

Volumetrics – It is difficult to obtain volumetrics for the genital regions. Yet, there still needs to be a method of objectifying the edema involvement. The HCP needs to capture the complete involvement with point-to-point girth measurements that can be repeated at future visits. One should look for bony landmarks (pubic bone, ischial tuberosity), anatomical structures (shaft of penis, labia, scrotum, anus), and/or scars or birthmarks.

Palpation – The HCP checks for pitting and/or underlying fibrosclerotic or fibroadipose tissue along the entire arm post breast cancer; the same practice should be followed with the genitals. Externally, pitting is commonly seen on the scrotum, labia, penis, vulvar vestibule, mons pubis, inner thigh, and gluteals/buttocks. Multiple locations on each structure may need to be evaluated. Internal evaluation for pitting of the vaginal tissue is possible and should only be conducted by pelvic floor therapists or other HCPs specifically trained in this form of assessment. The best place to start to "break the ice" in assessment of genital lymphedema for both the patient and the HCP is the trunk (lower abdomen and gluteals/buttocks) and then progress to the area just below the transverse perineal muscle. This area is prone to fluid accumulation and can provide evidence of possible genital involvement. To find the area, with the patient in a supine hook lying position the HCP needs to palpate the ischial tuberosity and then move the one finger medial ~ 1-2 inches and superiorly ~ ½ inch

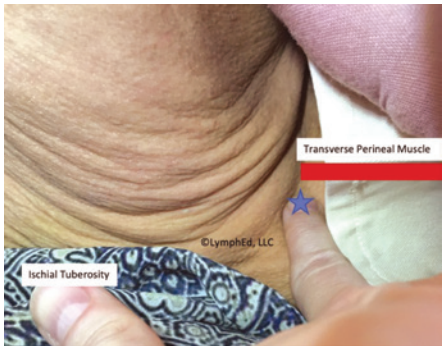
(please refer to image 2 on the right). This can be completed with the underwear still on, to help maintain modesty and to allow the patient to accommodate palpation of the genitals. This is especially key with females, as edema in this area can be the indicator of possible vaginal involvement without completing an internal evaluation.

Strength – Due to the deep location of the pelvic lymph nodes the muscle pump of surrounding muscles is vital for lymphatic drainage. The key muscles in the region include the transverse abdominals (TA), the pelvic floor muscles (PFM), the inner and outer thigh muscles, and the gluteals. Contraction of the pelvic floor muscles, or a Kegel, can be assessed in the same location as previously mentioned near the ischial tuberosity.

How is treatment amended for this population?

The genitals are prone to quick edema accumulation or refill due to the increased elasticity and the impact of gravity on the genital tissues as compared to other regions. The genitals are also susceptible to progression of tissue changes to adipose deposits and/or fibrosis, so early intervention is crucial for successful non-invasive treatment of the genitals.[4,7,8] The HCP should reassess the genital region in individuals with isolation of lymphedema to the leg(s) at initiation of each treatment, for it is not uncommon for progression to genital involvement.[21] To limit this risk, it is recommended to address the lower abdomen and genital region with compression (compression shorts) and manual lymphatic drainage (MLD) techniques in conjunction with lower extremity treatment.

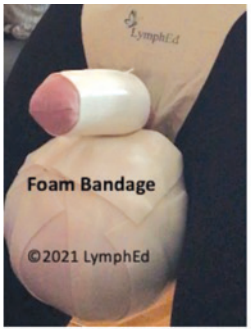
IMAGE 2

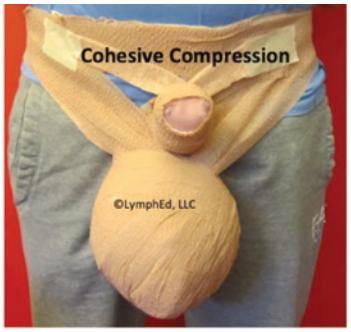


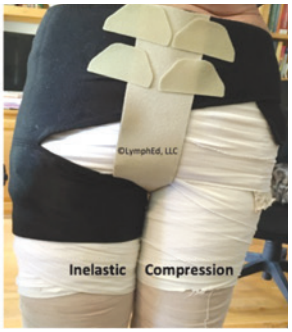
This image shows the location for assessing pitting. With the patient in hooklying position, locate the ischial tuberosity (thumb) and then move medially and superior to just below the transverse perineal muscle (just superior to index finger) to assess for pitting and for pelvic floor muscle contraction. You are directly on top of the fatty layer/tissue covering the levator ani pelvic floor muscles.

Compression – This is key to success with conservative and surgical interventions. The HCP must find a way to provide adequate compression and support to the region to limit edema and tissue changes. In both the treatment and maintenance phase of treatment, 23 hours a day compression/support needs to be implemented. It is common to need multiple additive components to obtain the desired results. For males this can include in the treatment phase, compression bandaging (elastic gauze, compressive foams) and support garments (jock straps, elastic shorts/underwear, support slings). Alternatively, one may use an all-in-one

IMAGES 3 - 4 - 5







These images show foam bandaging and cohesive compression on the penis and scrotum with the use of detachable genital models over clothing. The third image is of a female post gynecological cancer with bilateral lower extremity, truncal, and genital involvement. She has short stretch bandaging on bilateral lymphedema from the toes to the top of the thighs, medium stretch thigh bandaging bilaterally, inelastic thigh piece to address lateral hip area, and an inelastic piece posterior to anterior, with a foam pack underneath, to address labial edema.

system with foam and cohesive compression attaching around the abdomen. Compression bandaging is more difficult with females, especially if the patient is mobile, since anchoring to the trunk is more on an angle and often limits ambulation and urination/defecation. Foam inserts with inelastic genital straps attached to the trunk and an overlying support garment (underwear and/or shorts) are typically the most successful. In the maintenance phase, day compression is more for support to prevent gravity-linked fluid accumulation and the night compression is more for the reductive portion of treatment. Both garments are typically custom-made, since day compression off-the-shelf garments do not contain enough compression in the "panty" section. A main factor to always consider with genital edema is that compression needs to be breathable. This warm moist environment is prone to infections and this risk increases when improper compression is applied. The compression will only be successful if the patient, with assistance from family or other HCPs if necessary, is able to frequently remove and reapply all or portions of the compression system, for urination/defecation and hygiene!

FIGURE 1

Key points for patients regarding pelvic hygiene

- **Bathing:** Avoid hot baths, bubble baths, fragrant soaps, drying soaps, and scrubbing or over-drying the genital area.
- **Powders:** No fragrances. Corn starch is safe. Recommend lotion to powder for chaffing.
- **Toilet paper:** Avoid ones that leave a residue to prevent irritation. You can test by wiping with the same pressure on your pants and see if residue is left behind. This residue can be like "fiberglass" in the genital area.
- **Recommended to use toilet paper cleansing wipes** or to rinse with cool water via a cup after using the restroom and then pat dry to help remove excess bacteria and debris.
- **Females. Tampons:** use only non-fragrant and the lowest possible absorbency. You should be changing them every 2 hours to help decrease the risk of infection.
- **Males and Females. Pads:** Use only non-deodorant. You should be changing them every 2 hours to help decrease infections. You want a brand that can "wick" the fluid away from the surface, and not keep the moisture near your skin.
- **Male and Female Cosmetic Cleaning:**
  - Don't douche unless instructed by your medical doctor. This decreases your "good" bacterial count needed to fight infections
  - Don't shave, wax, laxer, pluck, dye, or pierce the genital area. Only trim the hair.
- **Infections:** If you think you have ANY infection, talk to your healthcare team. Signs: increased drainage, discharge, change in odor, increased itching/burning, increased pain, discomfort with urination, increased discoloration, and any change in your typical presentation.
- **Self-treatments for itching/burning symptoms:** Rinsing frequently with cool water, use a cool (not cold) pack – frozen peas wrapped in a dish towel.

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MLD – HCPs need to perform MLD on the trunk (especially the lower abdomen), and the genital area PRIOR to progressing into the lower extremities. If these regions are not fully addressed first, the lymphatic fluid moved up from the leg(s) could displace down into the genitals.[21] Previous research demonstrated the benefit of incorporating muscle contractions during MLD on the movement of fluid in the desired direction.[22-25] For genital and/or lower extremity involvement it is beneficial to incorporate the trunk muscles (specifically the TA and PFM) frequently during clinician led MLD and self-MLD. Due to adipose accumulation at the mons pubis in both sexes, posterior MLD pathways are typically more successful than anterior. Self-MLD at home needs to begin within the first few therapy sessions to improve results and to foster compliance with the home program post discharge. TA and PFM contractions should be a part of the self-MLD and prescribed exercises, with a minimal goal of 6-10 sets of these isometrics per day (the hold and repetitions to be determined by a HCP based on strength/endurance testing).

Education

Skin Care – Hygiene and skin care are crucial with genital lymphedema due to the fragility of the tissues leading to increased risk of skin breakdown/wounds and frequent infections/cellulitis.[2,4,9-11] The HCP needs to educate the patient (family and other HCP's, as appropriate) and assess for understanding on all aspects of skin care and prevention. The patient will typically need a handheld mirror to complete skin assessment at home.

Sexuality – This is the most overlooked portion of treatment. A main goal for HCPs in all diagnoses is a return to normal function, and

FIGURE 2

Key points for intimacy

- **Intimacy should NEVER hurt** for either males or females. Please speak to your medical professionals if you experience pain during or post intimacy.
- **Lubricants:**
  - Do not use Vaseline, baby oil, or Crisco.
  - Use only water-based products, avoiding multiple additives for they can produce irritation.
  - Safe substitutes: plain lubrication gels and olive oil
- **Hygiene:**
  - ALWAYS urinate after intimacy to decrease urinary tract infections.
  - Recommended to use a cleansing wipe or cool water to clean the area after intimacy to remove any irritants.
  - Perform your self-MLD as part of foreplay and post intimacy to encourage lymphatic fluid movement away from the genitals.
- **Positions:**
  - Avoid positions that compress or irritate the swollen tissues
  - Avoid positions where gravity can pull down on your swollen tissues
  - Males with swollen genitals typically have less worsening of swelling with sitting face-to-face with partner, kneeling with pillow supporting scrotum underneath, or missionary with scrotum supported on the bed.
  - Females with swollen genitals typically have less worsening of swelling by laying on their back with legs elevated and partner standing or kneeling, sitting face-to-face with partner, or sitting on top of partner where depth can be controlled to reduce friction on the swollen tissues.

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intimacy is a basic function. Intimacy does not always mean intercourse; it means to be close or familiar with someone, a connection. The psychosocial and physical symptoms associated with genital lymphedema (including odor, leakage, disfigurement), frequently limit this relationship or bond with others. A HCP needs to discuss the symptoms hindering intimacy and assist the patient in reducing the symptoms to return to a normal or an altered intimacy. The education can include altered positions for intercourse to limit pain or gravity/compression on the edematous tissue, methods to achieve orgasm without penetration, how to incorporate self/partner MLD as pre and post activities with intimacy, hygiene pre/post, and ways to alter previous intimacy (locating other erogenous areas, finding other activities to bring couples close, other types of intimacy). The HCP should openly discuss the topic and provide education or resources for the patient. It is not recommended for the HCP to allow the patient to perform or experiment with any forms of intimacy in the clinical setting! A referral to a psychologist or other HCP specializing in this area may be appropriate. The key is to open the dialogue and provide education or additional support services as needed. The therapist cannot assume that another HCP is addressing this issue.

Do you have any further messages for the HCP?

Early detection and treatment of the genital region is key for successful outcomes. HCPs cannot assume another HCP will address this taboo area nor that the edema will reduce only by addressing surrounding anatomic areas. An introductory subjective tool can assist in initiating the conversation between patient and HCP. All aspects of CDT are vital to the treatment of genital involvement. Including education on intimacy. HCPs and patients need to work together to remove the stigma associated with genital lymphedema. LP

A full set of references can be found at www.lymphedemapathways.ca

Editor's Note:
The author of this article is available to assist any HCP with evaluation or treatment questions via email at ShelleyDiCecco@LymphEd.com. The author is conducting a study pertaining to the education on and comfort level with treating genital lymphedema. HCPs may participate in the study by scanning the following QR image.

SCAN ME