LYMPHEDEMA (SWELLING) OF THE HANDS AND FEET

Swelling, Lymphoedema, Lymphatic Edema

An impaired lymphatic return and swelling of the hands and feet.

♀ Symptom ♂ Male & Female

About Lymphedema (swelling) of the hands and feet

Lymphedema is a disorder of the lymphatic vascular system characterized by chronic swelling of the affected extremity, recurrent infections, limited mobility and decreased quality of life.

The pathophysiology of lymphedema is generally divided into two periods. During the first period, the pathological changes occur mainly only in the lymphatics and in the soft tissues lymphedema symptoms are not apparent (occult lymphedema = Stage 0). After this stage, the pathological changes occur in the soft tissue (fat, connective tissue, skin, etc.) of the limbs, resulting in the progressive swelling (Pic. 1) caused by systematic and combined pathologic factors. This clinical state is characterized not only by progressive swelling but also by fat and scar deposition, immunosuppression (inhibiton of the immune response), a propensity for cellulitis, and microvascular proliferation (a hallmark of the cancer that begins in the brain).

As lymphedema worsens, a decrease of swelling after limb elevation of the limb (Stage 1; Pic. 2) will be not seen (Stage 2), and subsequently, the edema changes from pitting edema into nonpitting edema (late Stage 2). In many cases, the symptoms of lymphedema may be resistant to most of the therapies during this late Stage 2. Furthermore, edema is irreversible, and sclerosis of the skin and subcutaneous tissue (elephantiasis) may be remarkable (Stage 3).

Based on pathogenesis, one distinguishes two types of lymphedema:

- primary (hereditary) - results from genetic damage
- secondary (acquired) - a consequence of lymphatic failure resulting from trauma, surgery (Pic. 3), radiotherapy, or parasitic infection

Within the interstitial space (the space between tissue cells) fluid is always present. The amount of fluid depends on two factors: the amount introduced into the interstitial space, and the amount removed from it. Fluid enters the space from arterioles and venules; some returns to the venules, and the remainder is taken up by the lymphatics. In the normal physiologic state, entrance and exit are approximately equal, so that tissues retain their usual morphologic appearance and function. Edema (swelling) develops when the volume of interstitial fluid increases, either from increased inflow or decreased outflow, or both. An imbalance between lymphatic flow and the capacity of lymphatic circulation (capillary filtration exceeding the capacity of lymphatic circulation) bring up to edema. The protein constituents of static lymph fluid may cause inflammation and subsequent tissue fibrosis (the development of fibrous connective tissue). In patients with lymphedema, the overlying skin may develop cobblestoning (Pic. 4), as well as a verrucous or mossy appearance.

Lymphedema can be surprisingly difficult to diagnose, especially in its early stages. Without a proper diagnosis (Pic. 5), therapy is often delayed, allowing secondary fibrosis and lipid deposition to take place.

Despite substantial advances in surgical and conservative techniques, therapeutic options for the management of lymphedema are limited. There is no cure for lymphedema at this time, but treatments to manage and reduce the swelling include physiotherapy, massage, and compression bandages, known as complex physical therapy. Early treatment often results in rapid clinical improvement and prevents progression to the chronic phase of the disease.
Turner syndrome (TS) affects about 1:2,500 female newborns and is due to partial or total loss of the second sex chromosome. In the newborn, the finding of low birth length, webbed neck and lymphedema of the dorsum of hands and feet also points to diagnosis of TS.

Three genes were confirmed as the cause of congenital lymphedema in TS patients:

- VEGFR-3 (familial Milroy lymphedema)
- FOXC2 (lymphedema-distichiasis syndrome)
- SOX18 (hypotrichosis-lymphedema-telangiectasia)

Find more about related issues

**Diagnoses**

**Turner syndrome**
Turner syndrome is a genetic disorder in which a female is partly or completely missing one X chromosome that results in ovarian dysgenesis.
Learn more at: [www.fertilitypedia.org/therapy/diag/turner-syndrome](http://www.fertilitypedia.org/therapy/diag/turner-syndrome)

**Pic. 1: Bilateral leg lymphedema**

**Pic. 2: Lymphedema stages**
Staging according to the “consensus document” of the International Society of Lymphology.

<table>
<thead>
<tr>
<th>Clinical stage</th>
<th>Evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Subclinical, with possible clinical evolution</td>
</tr>
<tr>
<td>I</td>
<td>Edema regressing with treatments with positive pitting test</td>
</tr>
<tr>
<td>II</td>
<td>Edema partially regressing with treatments with negative pitting test</td>
</tr>
<tr>
<td>III</td>
<td>Elephantiasis with cutaneous complications and recurrent infections</td>
</tr>
</tbody>
</table>

**Pic. 3: Secondary lymphedema of the left arm after breast cancer surgery, including axillary lymphadenectomy**
Pic. 4: Cobblestone appearance of skin
African Elephant Rough Skin.

Pic. 5: The Stemmer-Kaposi sign
In lymphedema, the skin over metatarsal-phalangeal joint of the second toe can not be pinched into a fold (left, positive sign). In the absence of lymphedema (right), the skin can be readily pinched into a fold (negative sign).

Sources
“Lymphedema and Therapeutic Lymphangiogenesis” [https://www.hindawi.com/journals/bmri/2013/804675/] —by Saito et al. licensed under CC BY 3.0


“Topical tacrolimus for the treatment of secondary lymphedema” [http://www.nature.com/articles/ncomms14345#additional-information] —by Gardenier et al. licensed under CC BY 4.0


“African Elephant Rough Skin” [https://www.flickr.com/photos/mdpetttl/2743229739] —by Pettitt licensed under CC BY 2.0