

## A Patient Guide to Lymphedema and Turner Syndrome

### What is the Lymphedema?

**Lymphedema is a condition in which swelling occurs because of damage to or problems with the lymphatic system. This swelling does not go down with time or elevation.** The lymphatic system is a part of the circulatory system that removes excess fluid and waste from the body's tissues, filters them through the lymph nodes, and returns the fluid to the bloodstream. The lymphatic system also maintains your body's fluid levels and supports your immune system. If this system is not functioning properly, the fluid cannot move throughout the system as it should, but instead collects and "pools" in certain areas. If left untreated, it can lead to hardening of the tissue as well as an increased risk of infection.

### Turner Syndrome and Lymphedema

Because the lymphatic system sometimes does not develop fully before birth, if you have TS, you might be at a higher risk of developing lymphedema.

One of the most common noticeable physical features of a newborn with TS is puffy hands and feet caused by the buildup of fluid in surrounding tissues before birth. This extra fluid can also collect around the neck, forming what is called a **cystic hygroma**. After birth, this fetal lymphedema can cause a girl with TS to have a thicker, shorter neck with a "webbed" appearance, a low hairline at the back of the neck, low set ears, and narrow fingernails and toenails that curve upwards (**nail dysplasia**). Lymphatic swelling can occur at various points in life, not just childhood. If you are a woman with TS, you should be aware of warning signs and talk with your doctor if you have any questions or concerns.



A newborn's puffy hand, caused by prenatal lymphedema.

### What are the Warning Signs?

If you have Turner syndrome, you may have other medical concerns that seem more serious. Because of this, it can be easy for you and your doctors to miss the warning signs of lymphedema. Ignoring these can lead to it worsening or creating further difficulties.

#### If you are experiencing any of these warning signs, you should consult your doctor or see a lymphedema specialist:

- Do you have a full or heavy feeling in your leg or arm?
- Are you unable to pinch skin up easily on top of your foot or hand?
- Do you feel a tightness in the skin or tissue of certain areas?
- Have you noticed decreased flexibility in your hand, foot, wrist, or ankle?
- Are you having difficulty fitting into clothing in a specific area?

**"My toddler kept having 'general infections' and would get a high fever and become lethargic. Eventually, she was diagnosed with cellulitis, an infection related to her lymphedema. The clue was that each time she had the infections, her foot was red and warm to the touch."**  
**-Mom of daughter with TS.**

**If you notice a rash, itching, redness, pain, increase of temperature, or fever, you should see your physician immediately.**

## Treatment for Lymphedema

Although there is no cure for lymphedema, therapy and treatment can bring a lot of relief, especially if it is caught in the earlier stages. The standard of treatment for lymphedema is called **Complete Decongestive Therapy**, which involves a gentle massage called Manual Lymphatic Drainage (MLD) and the use of compression garments and bandaging to reduce and maintain the size of the limb.

## What to Expect During Your Therapy

Treatment will often include an office visit, where your therapist will massage, and help with drainage before wrapping the limb with compression bandages. You will be asked to go home and come back at a later time to have the bandages redone. Depending on the amount of swelling, this might take a few trips. It is very important that you **DO NOT** attempt to wrap your own limbs with the compression bandages. Doing so can cause increased damage.

Once the swollen limb is back to normal size, your therapist will prescribe wearing compression garments to maintain the reduced swelling.



Compression garments are used to reduce swelling and maintain the size of the limb.

## A Word About Infection

There are numerous reasons why patients with lymphedema are at an increased risk for infections. Normally, the body is protected by a fine acid layer on the surface of the skin, which prevents bacteria and other pathogens from entering. However, the skin in lymphedema tends to be dry and scaly, causing a disruption of the protective acid layer. If deepened skin folds are present, moisture collecting in these folds may create a breeding ground for bacteria.

## Lymphedema Fast Facts:

- Lymphedema is most commonly found in TS patients with a non-mosaic 45, X karyotype, and usually resolves by 2 years of age, although it can persist throughout life and recur.
- In a lymphedema-focused survey (1), 65% of girls and women with a 45,X karyotype experienced lymphedema.
- The most common treatment for lymphedema is Complete Decongestive Therapy
- If left untreated, lymphedema can lead to hardening of tissue and increased risk of developing an infection, such as cellulitis.
- There is no cure for lymphedema, but with treatment and therapy, there are ways to control the symptoms.

## For More Information, Contact Us: Turner Syndrome Society of the US

11250 West Road, Suite G | Houston, TX  
77065

1.800.365.9944 | E-mail:  
tssus@turnersyndrome.org  
www.turnersyndrome.org

Visit [www.lymphnet.org](http://www.lymphnet.org) to learn more about lymphedema.

### References:

1. Rothbauer, J, Driver, S, Callender, L. Describing Lymphedema in Females with Turner Syndrome. *Lymphology* 2015 **48** 139-152
2. Bondy, CA. Care of girls and women with Turner syndrome: a guideline of the Turner syndrome study group. *J Clin Endocrine Metab* 2007 **92**