Overview of Vascular System

• The vascular system (also known as the circulatory system) is comprised of vessels that transport blood and lymph through the body
• Veins and arteries carry blood through the body, deliver oxygen and nutrients to the body tissues and remove tissue waste matter
Overview of the Vascular System

- The vessels of the blood circulatory system are:
  - Arteries: carry oxygenated blood away from the heart to the body
  - Veins: carry blood from the body back to the heart
  - Capillaries: tiny blood vessels between arteries and veins that distribute oxygen-rich blood to the body

What is a Vascular Anomaly

- Occurs when the tubes that carry blood or lymph do not develop properly
- Many different conditions
- May be present at birth or develop soon after
Types of Vascular Anomalies

- Vascular Tumors: hemangioma, pyogenic granuloma, glomangioma, Kaposi’s sarcoma, angiosarcoma, hemangioendotheliomas

- Vascular Malformations: capillary vascular malformations (port-wine stains), venous malformation (abnormal veins), arteriovenous malformation (abnormal arteries and veins), spider angiomas (arteries that show up on the surface of the skin with a centralized dot and outer branches), angiofibromas (pink raised skin bumps seen in patients with tuberous sclerosis), lymphatic malformations (abnormal lymphatics or lymph vessels which can lead to enlarged water-filled cysts and swelling)
Types of Vascular Anomalies Continued

• Complex combined malformations (sometimes accompanying a genetic syndrome), including:
  – Sturge-Weber syndrome (port wine stain in the upper third of the face, with eye and central nervous system involvement)
  – Klippel-Trenaunay syndrome (abnormal capillaries, veins, and lymphatics with overgrowth of the involved area of the body)
  – Parkes-Weber syndrome (port wine stain with arteriovenous malformation and overgrowth of the involved area of the body)
  – Hereditary hemorrhagic telangiectasia (may have nose and GI bleeding from small dilated vessels)
  – CLOVES syndrome (a collection of conditions that may include congenital lipomatous overgrowth, vascular malformations, epidermal moles, and spinal abnormalities)
  – Proteus syndrome (vascular malformations and overgrowth of body parts)

Medical management of vascular anomalies

• Interventional radiology: sclerosing injections
• Medications (hemangioma)
• Laser treatment
• Monitoring
• Surgical Intervention
• Compression Garments
Overview of Lymphatic System

- A system of lymphatic capillaries, vessels, nodes and ducts that collect and transport lymph
- Lymph is a clear fluid, similar to plasma in blood
- Lymphatic system is important for maintaining your body’s fluid balance and works with the rest of the immune system to fight infections
Overview of Lymphatic System Continued

- Lymph does not circulate in the body the way that blood does
- Lymph vessels are both superficial and deep
- Lymphatic vessels are found in most organs and tissues of body
- Lymphatic vessels carry the lymph to lymph nodes
- Lymph nodes are mostly found around the neck, armpits, groin, thorax, knees and elbow
- Lymph nodes work to filter and monitor the lymph for foreign particles such as pathogens and cancerous cells before it returns to the blood

Overview of Lymphatic System Continued

- After lymph is filtered, it leaves the node and travels to lymphatic trunks and lymphatic ducts
- After reaching the lymphatic ducts, it then joins the blood in the venous system

http://www.bing.com/videos/search?q=lymphedema+video+for+kids&view=detail&mid=B466EDFDCEAD69ACE5FDB466EDFDCEAD69ACE5FD&FORM=VIRE&adlt=strict
What is a lymphatic malformation or “lymphedema”

- Lymphedema is an abnormal accumulation of protein rich fluid (lymph), usually in the extremities due to insufficient lymphatic circulation, causing limb swelling, reactive fibrosis, and chronic inflammation.
- Can be caused by infections, cancer, surgical removal of lymph nodes, damage of lymph nodes from radiation therapy, vessels formed improperly, or inherited conditions.
Staging of Lymphedema

**Primary vs. secondary lymphedema**

- **Primary Lymphedema**: a developmental abnormality of the lymphatic system

- **Secondary Lymphedema**: caused by a known insult to the lymphatic system
Primary Lymphedema

- Congenital or hereditary
- Can present as a variety of abnormalities
- Classified by the age of the patient at the onset of swelling
- Almost exclusively affects the lower extremity (unilaterally or bilaterally) but can affect upper extremities as well
- Greater prevalence in females
- Swelling starts distally and moves proximally
- Occurs without known impetus or after minor trauma (insect bites, injections, sprains, strains, burns, cuts), infections, or immobility
- Triggers produce additional stress to the already impaired lymphatic system which leads to a mechanical insufficiency

3 Most Common Forms of Primary Lymphedema

- **Congenital lymphedema / Milroy’s disease**
  - Clinically evident at birth or within the first 2 years of life
  - Familial pattern of inheritance

- **Lymphedema praecox / Meige’s disease**
  - After birth but before the age of 35
  - Most common form
  - Most often arises during puberty or pregnancy

- **Lymphedema tarda**
  - After age 35

- **Associated Conditions**
  - Amniotic Band syndrome
  - Turner’s syndrome
  - Noonan’s syndrome
  - Klippel-Trenaunay syndrome
  - Parkes Weber syndrome
Secondary Lymphedema

• In pediatrics, the most common cause of secondary lymphedema is radiation treatment of cancer

• Other causes include surgery, trauma, infection, malignant tumors, immobility, and chronic venous insufficiency
Specific Risk Factors in Cancer Treatment

- Radiation to the following areas produces the greatest risk for development of lymphedema: underarm area, groin, pelvis, head/neck
- Surgical removal of lymph nodes (the greater the number of nodes removed, the greater the risk for lymphedema development)
- Scar tissue in the lymphatic ducts or veins, under the collar bones, caused by surgery or radiation therapy
- Cancer that has spread to the lymph nodes in the neck, chest, underarm, pelvis, or abdomen
- Tumors growing in the pelvis or abdomen that involve or put pressure on the lymphatic vessels and/or the large lymphatic duct in the chest and block lymph drainage
- An inadequate diet or those who are overweight. These conditions may delay recovery and increase the risk for lymphedema.

Incidence/Prevalence

- WHO estimates that over 150 million people worldwide have secondary lymphedema
- NIH states that the incidence of primary lymphedema could be as high as 1 in 300 live births
- Of the 1.4 billion people in 73 countries threatened by lymphatic filariasis, commonly known as elephantiasis, 120 million are currently infected and 40 million of these patients are disfigured and incapacitated by the disease
- CDC warns that the incidence of secondary lymphedema among cancer survivors is between 20–40%
Medical management of lymphedema

- Diagnostics (MRI, lymphangintigrophy)
- Genetics (can test for certain types of primary lymphedema (Meige Disease, Milroy’s Disease, Noonan’s, etc.)
- Consultation with Vascular Medicine and/or Dermatology to maintain skin integrity and prevent infections (cellulitis)
Therapeutic management

- Complete Decongestive Therapy (CDT)
  - Manual Lymphatic Drainage: a gentle manual treatment technique with the goal of rerouting lymph flow around blocked areas into more centrally located healthy lymph vessels, which drain into the venous system
  - Compression Therapy: begins with intensive bandaging phase using short stretch bandages and progresses to daytime/nighttime compression garments with the goal of aiding fluid return, maintaining the decongestive effect achieved during MLD, and preventing re-accumulation of fluid into the tissues

- Manual Lymph Drainage
  - https://www.youtube.com/watch?v=dT6rAL4-D14

- Compression Bandaging
  - https://www.youtube.com/watch?v=f3UAjU2-sAE
Therapeutic management

• Home Exercise Program
  - Goal is assist the body with remodeling the tissue and decongesting the affected tissue by increasing the return of lymphatic fluid to the circulatory system
  - Can be physical activity as well as diaphragmatic breathing
  - Exercises need to be started slowly and progressed gradually while monitoring for any significant changes
  - Exercises should be completed with compression garments donned for greatest benefit
Therapeutic management

- **Family education**
  - Educate family/patient prior to radiation therapy/lymph node removal on potential risk of lymphedema development
  - Educate on what signs/symptoms to look for
  - Address proper skin care and grooming of affected area
  - Ensure caregivers understand and can demonstrate proper MLD techniques, compression bandaging techniques and donning/doffing of compression garments

Common insurance challenges with management of lymphedema

- **Most insurance companies do not cover the cost of bandages for compression wrapping**
  - Supplies for bandaging can cost approximately $100 per month
- **Many insurance companies do not cover compression garments, or only cover a limited amount per year**
- **Many insurance companies will not cover the cost of a custom-sized compression garment**
  - A set of custom compression garments can cost over $1,000 out of pocket
The caregiver experience: Katie

Case Study
Case Study

• Born 39 weeks with no complications
• 3 older brothers and sisters with no family hx of lymphedema
• Parents noticed swelling of UE’s at birth; officially diagnosed with primary lymphedema at 7 months of age

• Began with 23 hr per day UE wrapping with compression bandages at very early age
• Other issues began to arise including: protein losing enteropathy, lymphangictasia, etc. Has to drink a special formula and eat a modified diet.
• Family moved from Portland, Oregon to Ohio when he was 16 months old
Case Study

• Family struggled to find a therapist who would bandage and get garments for a young child
• Family fought to get any coverage for custom sized compression garments
• He continued to be bandaged for 23 hrs per day which was developing into tactile defensiveness on UE’s and limiting his development of fine motor, play and self-care skills
• Obtained custom sized compression gloves- requires 2 pairs at a time due to wash/wear and needs new garments every 4-6 months.
• Was able to start doing more with his hands and used time out of gloves for lots of tactile play
• Still follows this schedule- wears compression gloves during day and is bandaged at night
Case Study

- Had Lymphoscintigraphy in June of 2014
- Results:
  - Upper extremities: bilateral delayed lymphatic flow. No significant activity identified in either axilla
  - Lower extremities: delayed lymphatic flow in the left lower extremity. Reasonably better drainage of the right lower extremity than left
- Got preventative compression stockings for lower extremities
- Has been seen by genetics with no significant findings
- Followed by dermatology, GI, pulmonology, allergy/immunology, genetics, vascular medicine and vascular anomalies clinic
- Gets OT 1x EOW; focus on tactile play, fine/visual motor, UE strengthening, compression garment management, caregiver education, home programming

What is the vascular anomalies clinic

- Specialists from several departments, including: dermatology, radiology, reconstructive surgery, genetics, pathology, hematology, ophthalmology, orthopaedics and therapy services
- The team meets regularly for case-discussions regarding our referral patients, as well as to discuss ongoing matters related to vascular anomalies.
- Clinic is held at Main Campus on the second Monday of each month
How to refer to vascular anomalies clinic

• **Call:** 216.445.4726 or 800.223.2273, ext. 54726

How to refer to Jenny Negrey for therapeutic management of vascular/lymphatic concerns

• E-mail directly at [negreyj@ccf.org](mailto:negreyj@ccf.org)
• Contact Denise Kaufman at TS-Westlake requesting OT services with Jenny Negrey for vascular/lymphatic concerns 440-835-7400
References


Thank you!

- Questions?