

Primary Lymphedema: Diagnostic Decision Tree

Simplified from the St George's Hospital Algorithm

START HERE: Is the swelling Primary or Secondary?

- **Rule out Secondary Causes first:** Has the patient had surgery, radiation, trauma, or a known infection (cellulitis)?
 - **If NO:** Proceed to the Primary Lymphedema screening below.
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1. What is the Age of Onset?

- **Birth to 1 Year (Congenital):**
 - *Check for:* Bilateral swelling, hydrocele (in males).
 - *Common Diagnosis:* Likely **Milroy Disease**.
 - **Puberty to Age 35 (Praecox):**
 - *Check for:* Distichiasis (double eyelashes), varicose veins, or cleft palate.
 - *Common Diagnosis:* Likely **Lymphedema-Distichiasis** or **Meige Disease**.
 - **After Age 35 (Tarda):**
 - *Action:* **High suspicion.** Rule out pelvic malignancy or venous obstruction before labeling as Primary.
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2. Are there Associated Features?

Follow the branch based on the "extra" symptoms beyond swelling:

- **Limb Swelling ONLY:**
 - Usually presents at puberty. No other health issues.
 - *Pathway:* Routine Lymphedema Management.
- **Swelling + Overgrowth:**
 - One limb is significantly longer or larger in bone/tissue structure.

- *Pathway:* Refer to Orthopedics and Genetics (possible PIK3CA-related overgrowth).
 - **Swelling + Systemic Issues:**
 - Patient has history of pleural effusion, pericardial effusion, or chronic diarrhea (protein loss).
 - *Pathway:* **Urgent Referral** to Internal Medicine/Genetics.
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3. The Physical Exam "Quick-Hitters"

If the doctor only has 60 seconds, they should check these three things:

1. **Stemmer's Sign:** Can you pinch the skin on the base of the second toe? (If **NO**, it is Lymphedema).
 2. **Toes:** Are they "squared off" or boxy looking?
 3. **Distribution:** Does the swelling start at the ankle (Venous) or the toes (Lymphedema)?
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Primary Care Action Plan

- **Education:** Focus on skin integrity to prevent cellulitis.
 - **Referral:** Direct to a Certified Lymphedema Therapist (CLT) for decongestive therapy.
 - **Avoid:** Diuretics are not indicated for Primary Lymphedema.
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This chart is a simplified tool for primary care screening. For the full clinical St George's Classification, visit