

A 1-day-old girl is in the newborn nursery with swollen hands and feet. She was born at term by uncomplicated vaginal delivery. Her mother had mild pre-eclampsia and swelling of her hands and feet during pregnancy. The pregnancy was otherwise uncomplicated. Vital signs are normal. Examination shows a girl with a short webbed neck, dysplastic nails, and bilateral, non-pitting carpal and pedal edema. Ultrasound of the abdomen shows a horseshoe kidney. Which of the following is the most likely cause of the edema?

- ☐ A. Decreased albumin synthesis
- ☐ B. Decreased cardiac contractility
- ☐ C. Dysgenesis of the lymphatic network
- ☐ D. Increased capillary permeability
- ☐ E. Increased sodium retention
- ☐ F. Increased urinary loss of protein

Submit



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- ☐ A. Decreased albumin synthesis [3%]
- ☐ B. Decreased cardiac contractility [5%]
- ☒ C. **Dysgenesis of the lymphatic network** [65%]
- ☐ D. Increased capillary permeability [5%]
- ☐ E. Increased sodium retention [10%]
- ☐ F. Increased urinary loss of protein [12%]

Proceed to Next Item

Explanation:

User Id: [REDACTED]

| Causes of edema                                |  |
|--|--|
| Primary mechanism                              | Examples   |
| ↑ Capillary hydrostatic pressure               | <ul style="list-style-type: none"> <li>Heart failure</li> <li>Glomerulonephritis, renal failure</li> <li>Venous obstruction (eg, cirrhosis, venous insufficiency)</li> </ul>           |
| ↓ Capillary oncotic pressure (hypoalbuminemia) | <ul style="list-style-type: none"> <li>Protein loss (eg, nephrotic syndrome, protein-losing enteropathy)</li> <li>Decreased albumin synthesis (eg, cirrhosis, malnutrition)</li> </ul> |
| ↑ Capillary permeability                       | <ul style="list-style-type: none"> <li>Burns, trauma &amp; sepsis</li> <li>Allergic reactions</li> </ul>   |



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| ↑ Capillary permeability                       | <ul style="list-style-type: none"> <li>Burns, trauma &amp; sepsis</li> <li>Allergic reactions</li> <li>Other systemic inflammatory processes</li> </ul>                                |
| Lymphatic obstruction                          | <ul style="list-style-type: none"> <li>Malignancy &amp; related treatment</li> <li>Hypothyroidism</li> <li>Congenital lymphedema</li> </ul>  |

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This girl's clinical presentation (eg, **webbed neck**, **carpal and pedal edema**, nail dysplasia, horseshoe kidney) are strongly suggestive of **Turner syndrome**. The edema is likely due to **congenital lymphedema** from abnormal development of the lymphatic network. The dysfunctional lymphatic system causes accumulation of protein-rich interstitial fluid in the hands, feet, and neck (webbed neck). Severe obstruction of lymphatic vessels can result in **cystic hygroma** of the neck and fetal hydrops.

Lymphedema is generally **nonpitting** as opposed to the pitting edema seen with liver failure (**Choice A**), congestive heart failure (**Choice B**), or nephrotic syndrome (**Choice F**). In addition, hepatomegaly is seen in right heart failure and liver failure, but not in lymphedema.

(**Choice D**) Pre-eclampsia/eclampsia can exacerbate extremity swelling in pregnancy



|                         |   |
|-------------------------|---|
| ↑Capillary permeability | <ul style="list-style-type: none"><li>• Burns, trauma &amp; sepsis</li><li>• Allergic reactions</li><li>• Other systemic inflammatory processes</li></ul> |
| Lymphatic obstruction   | <ul style="list-style-type: none"><li>• Malignancy &amp; related treatment</li><li>• Hypothyroidism</li><li>• Congenital lymphedema</li></ul>             |

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(**Choice D**) Pre-eclampsia/eclampsia can exacerbate extremity swelling in pregnancy due to increased capillary permeability, decreased albumin, and renal dysfunction. These faulty mechanisms are not the primary problem in congenital lymphedema.

(**Choice E**) Increased sodium and fluid retention is seen in normal pregnancy (total body volume expansion) as well as renal pathology and heart failure.

**Educational objective:**

Turner syndrome should be suspected in newborn girls who have a webbed neck, horseshoe kidney, carpal and pedal edema, and nail dysplasia. Congenital lymphedema occurs due to abnormal development of the lymphatic system.

**References:**

1. [The role of lymphoscintigraphy in the diagnosis of lymphedema in Turner syndrome.](#)
2. [Lymphedema: a primer on the identification and management of a chronic condition in oncologic treatment.](#)



Media Exhibit

syndrome

