

A newborn boy is evaluated shortly after vaginal delivery for respiratory distress. He was born at 37 weeks gestation to a primigravid mother who had no medical problems. Second-trimester prenatal ultrasound showed bilateral hydronephrosis and oligohydramnios. His temperature is 36.7 C (98 F), pulse is 144/min, and respirations are 50/min. Pulse oximetry is 84% on room air. Examination shows decreased aeration of both lungs and intercostal retractions. The child has flattened facies, abdominal distension, and clubfeet. What is the most likely underlying diagnosis in this patient?

- ☐ A. Bilateral cryptorchidism
- ☐ B. Congenital diaphragmatic hernia
- ☐ C. Duodenal atresia
- ☐ D. Hypospadias
- ☐ E. Posterior urethral valves
- ☐ F. Prostatic hyperplasia
- ☐ G. Wilms tumor

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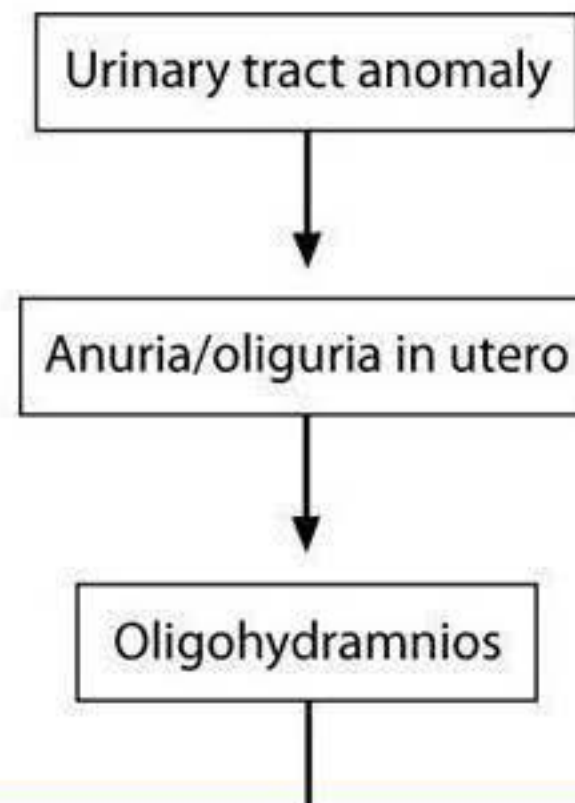
- ☐ A. Bilateral cryptorchidism [1%]
- ☐ B. Congenital diaphragmatic hernia [9%]
- ☐ C. Duodenal atresia [8%]
- ☐ D. Hypospadias [2%]
- ☒ E. **Posterior urethral valves** [71%]
- ☐ F. Prostatic hyperplasia [1%]
- ☐ G. Wilms tumor [8%]

Proceed to Next Item

Explanation:

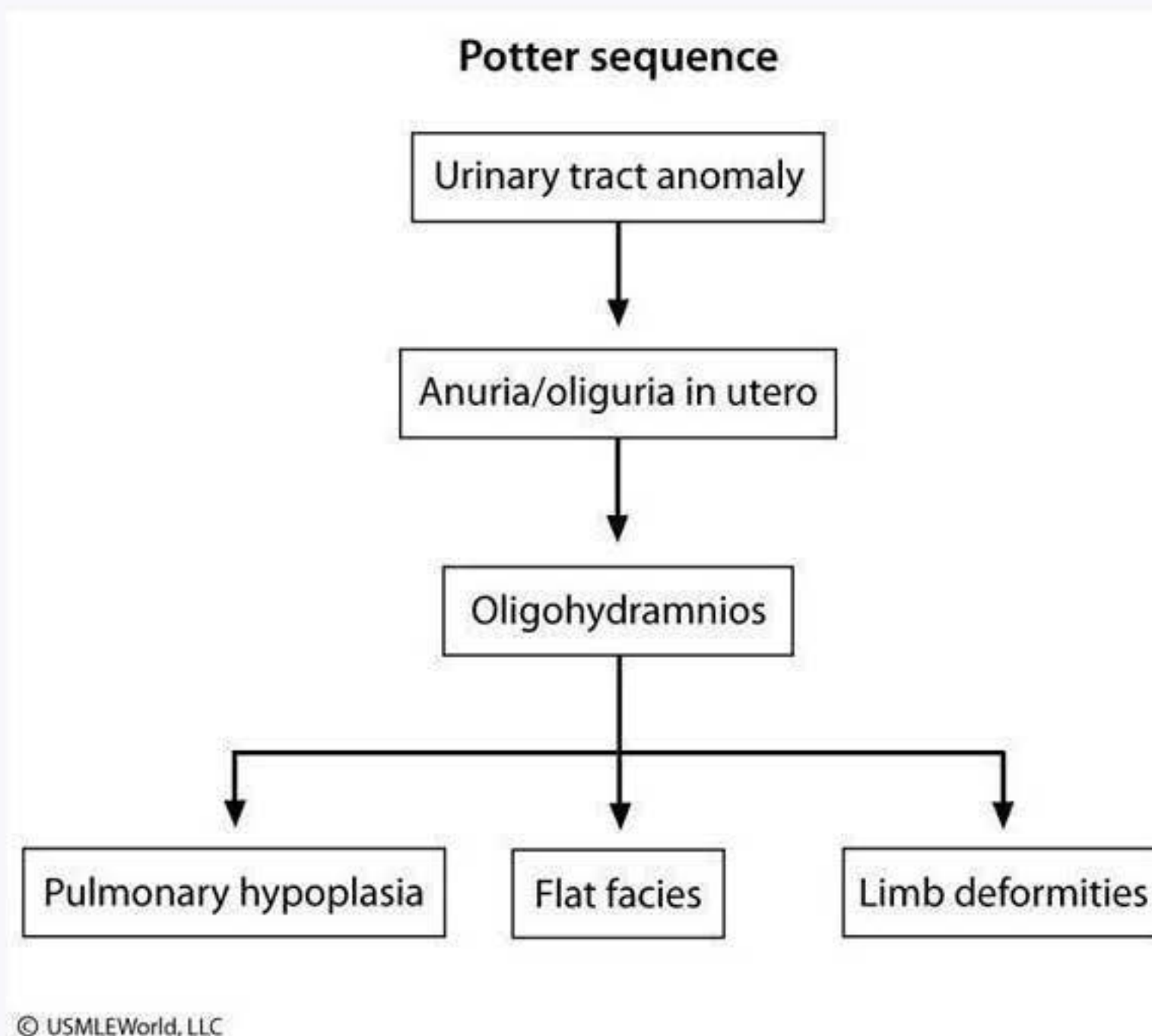
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Potter sequence



Explanation:

User Id:



Posterior urethral valves (PUV) are the most common cause of urinary tract obstruction in newborn boys. Abnormal folds in the distal prostatic urethra obstruct urinary flow, resulting in **progressive dilation of the bladder, ureters and kidneys**. Prenatal ultrasonography findings of **bladder distention, bilateral hydroureters, and bilateral hydronephrosis** are highly suggestive of PUV. Poor urine output in utero results in oligohydramnios as fetal urine is a major source of amniotic fluid. **Oligohydramnios** in the second trimester is ominous and associated with high perinatal mortality because normal amniotic fluid levels are required for lung development. Low amniotic fluid also restricts fetal movement, leading to a cascade of physical anomalies including flat facies and limb deformities (**Potter sequence**).

Other affected infants can present with poor urinary stream, straining with voiding, urosepsis, failure to thrive, and **renal failure**. Diagnosis is confirmed by voiding

Pulmonary hypoplasia

Flat facies

Limb deformities

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Other affected infants can present with poor urinary stream, straining with voiding, urosepsis, failure to thrive, and **renal failure**. Diagnosis is confirmed by voiding cystourethrogram and cystoscopy. Treatment options include PUV ablation and urinary diversion. Despite prenatal diagnosis and early surgical intervention, patients are at high risk for permanent kidney damage.

(Choice A) Cryptorchidism is the failure of one or both testes to descend from the abdomen through the inguinal canal(s) into the scrotum. Examination shows a hypoplastic or poorly rugated scrotum. However, cryptorchidism would not cause obstructive uropathy and oligohydramnios.

(Choice B) Congenital diaphragmatic hernia is a life-threatening defect of the diaphragm that allows abdominal viscera to herniate into the chest, resulting in restriction of lung development. Patients can present with respiratory distress, but on examination the abdomen appears scaphoid due to loss of the abdominal contents into the chest. Prenatal ultrasound demonstrates a mass in the chest (abdominal contents) and sometimes polyhydramnios from esophageal compression, making this diagnosis unlikely.

(Choice C) Fetal swallowing is responsible for amniotic fluid removal. An atretic duodenum can obstruct amniotic fluid clearance, resulting in polyhydramnios, in contrast to the oligohydramnios seen in this patient. Another significant finding on prenatal ultrasound includes the characteristic "double bubble" sign due to dilation of the stomach and the first part of the duodenum.

(Choice D) Hypospadias is a congenital abnormality in which the penile urethra opens on the ventral surface of the penis rather than the tip. These patients are able to urinate but may have difficulty controlling the urinary stream.

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(Choice D) Hypospadias is a congenital abnormality in which the penile urethra opens on the ventral surface of the penis rather than the tip. These patients are able to urinate but may have difficulty controlling the urinary stream.

(Choice F) Benign prostatic hyperplasia can cause obstructive uropathy at the same level as posterior urethral valves. However, this condition is common in men age >50 and does not affect children.

(Choice G) Wilms tumor (nephroblastoma) is the most common renal malignancy in childhood. Most children present at age 2-5 years with a large, palpable flank mass but no other symptoms. It is extremely rare for Wilms tumor to present in utero.

Educational objective:

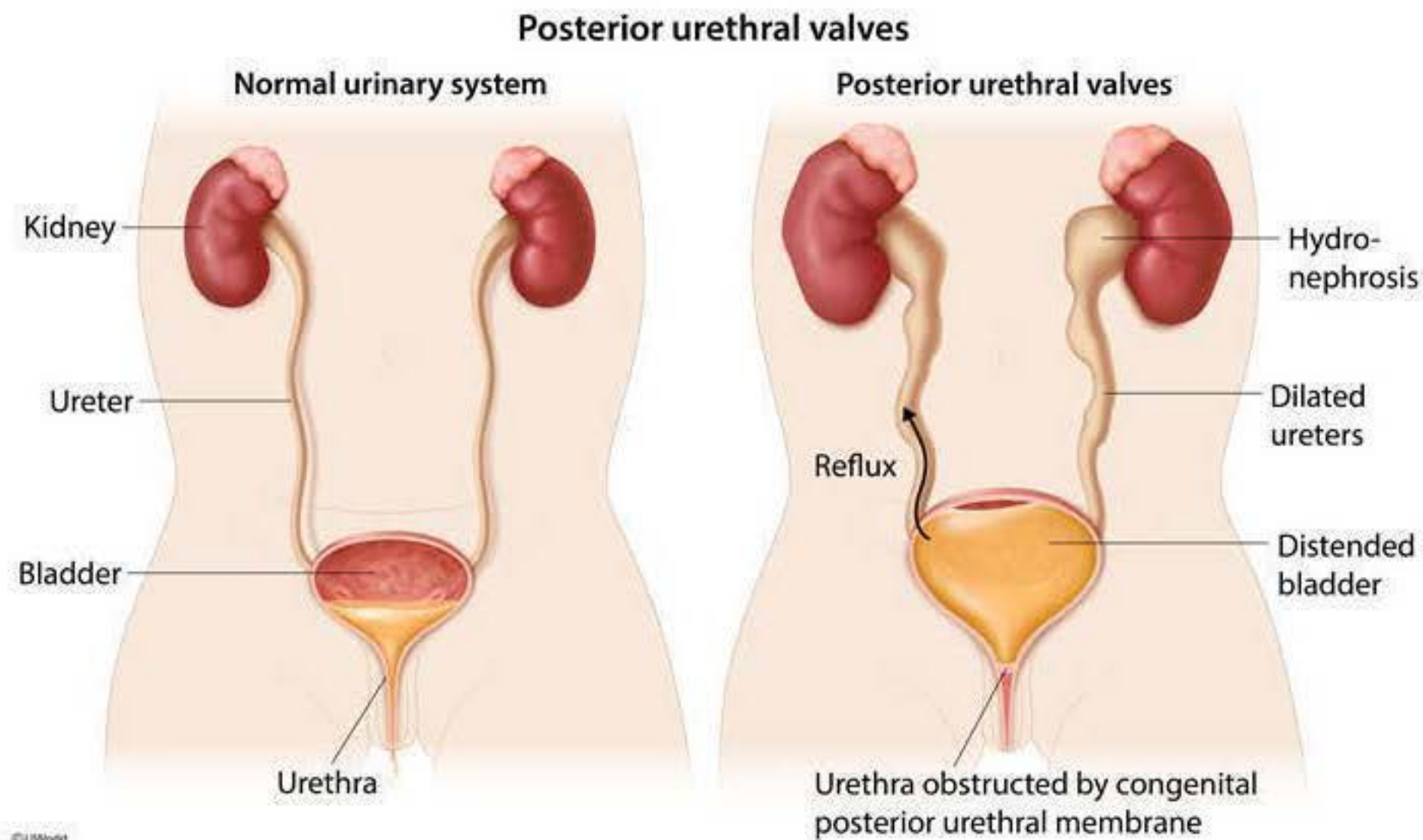
Posterior urethral valves are the most common cause of urinary tract obstruction in newborn boys. Classic findings on prenatal ultrasonography include bladder distension, bilateral hydroureters, and bilateral hydronephrosis. Oligohydramnios from low urine production in utero can cause pulmonary hypoplasia and postnatal respiratory distress.

References:

1. [Congenital urethral anomalies in boys. Part I: posterior urethral valves.](#)
2. [Risk factors for end stage renal disease in children with posterior urethral valves.](#)
3. [Fetal surgery for posterior urethral valves: long-term postnatal outcomes.](#)

Media Exhibit

or urethral valves



Media Exhibit

orchidism

