Congenital Posterior Urethral Valves (PUV)

What is it?

Congenital posterior urethral valves (PUV) is a disorder that affects the male genitourinary system. It occurs when extra tissue grows inside of the **urethra** and interrupts the flow of urine. The urethra is a small tube connected to the bladder that delivers urine to the outside of the body. The kidneys, bladder, ureters, and urethra will dilate or swell if the urine cannot exit the body. PUV can be found during pregnancy by ultrasound. It can range from mild to severe depending on how much urine is blocked. Urine blockage can cause severe damage to the genitourinary organs. If the PUV case is mild, a boy may not experience any symptoms until age 10 or beyond. Mild symptoms may include urinary tract infection, painful urination, bedwetting, a weak urine stream, and an enlarged bladder. Severe symptoms can include respiratory distress, bladder dysfunction, vesicoureteral reflux, and **hydronephrosis**. Hydronephrosis occurs when too much urine accumulates inside the kidneys, leading to swelling and pain. It is essential to maintain proper bladder function for boys with PUV because they are at an increased risk of developing kidney failure.

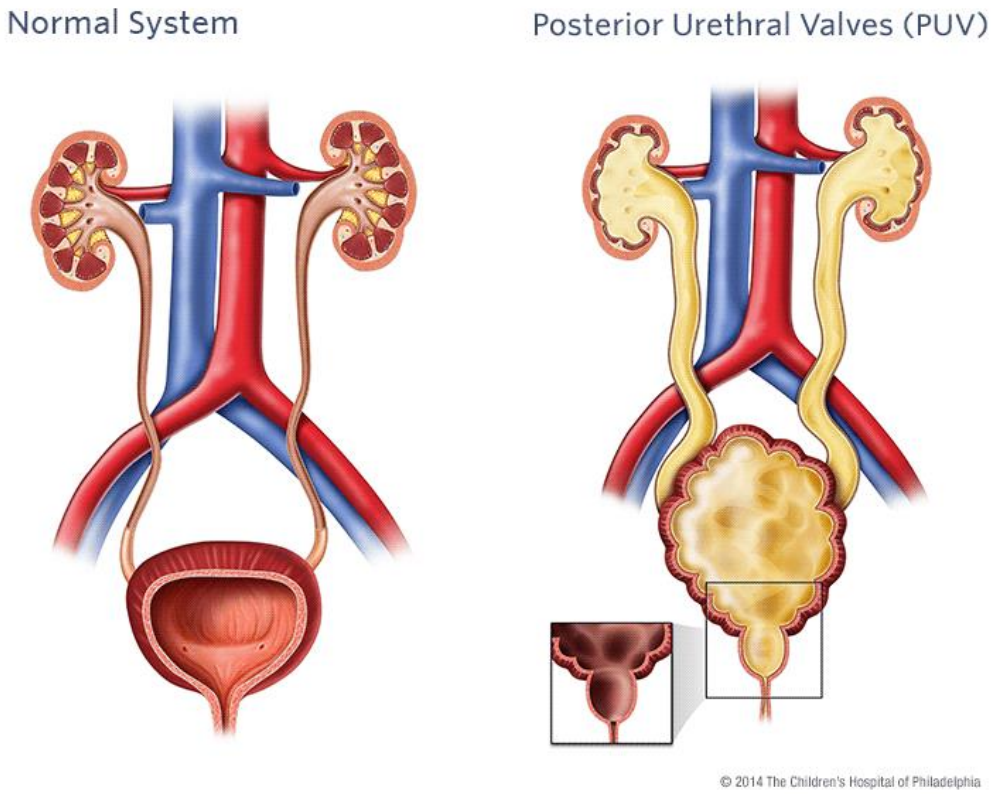

Image courtesy of the Children's Hospital of Philadelphia

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How common is it?

PUV occurs only in boys. It is estimated to be found in about 1 in every 8,000 births.

What causes it?

The exact cause of posterior urethral valves is unknown. There may be many factors that cause it, including a possible genetic component. It is thought to occur during the early fetal development of boys.

How is it diagnosed?

It can be identified during pregnancy with an ultrasound but cannot be officially diagnosed until birth. Once the baby is born, a diagnosis can be made by performing a urodynamic test (looks inside the bladder), a cystourethroscopy (looks inside the urethra), blood tests, or avoiding cystourethrogram (x-ray of the entire urinary tract).

How is it treated?

In most cases, PUV can be treated with surgery called endoscopic urethral valve ablation. An instrument is used to break down the valves to remove the tissue obstruction inside the urethra. A vesicostomy procedure may be performed if the tissue obstruction is too severe. A small opening is made in the bladder to allow the urine to empty. This opening is temporary and will eventually require closure. The genitourinary system will need to be monitored throughout the child's lifetime to ensure proper function.

For more information:

Children's Hospital of Philadelphia
https://www.chop.edu/conditions-diseases/posterior-urethral-valves-puv

Boston Children's Hospital
https://www.childrenshospital.org/conditions-and-treatments/conditions/p/posterior-urethral-valves

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