Posterior Urethral Valves

What are Posterior Urethral Valves?
The urethra is a tube that carries urine from the bladder to outside the body. Posterior urethral valves (PUV) are abnormal congenital (a condition present or recognized at birth) valves that block the urinary flow in the urethra. Valves are flaps of flesh which attach to the walls of the urethra and partially block the flow of urine, much like spinnakers on a sailboat block the flow of the wind.

How can PUV cause problems?
Posterior urethral valves can cause a variety of problems. The bladder wall muscle can thicken and enlarge from the extra work involved in voiding (bladder hypertrophy), like the muscles of a body builder with exercise. Unlike the bodybuilder, the bladder may enlarge and stretch to the point the muscles become over stretched and too weak to work. This condition may require intermittent catheterization to empty the bladder. In addition, valves may result in vesicoureteral reflux, the movement of urine backwards from the bladder back to the kidney. The result of reflux is dilation of the ureters (the tube that connects the bladder to the kidney) and the kidney itself. This predisposes the kidney to infections and kidney damage. Renal dysplasia, which is the abnormal development of the kidney, occurs in association with posterior urethral valves. Dysplastic kidneys function poorly or not at all.

Severe cases of posterior urethral valves may result in life-threatening problems. Decreased urine output from the fetus results in decreased fluid in the amniotic sac (fluid in the sac is fetal urine). This condition, called...
oligohydramnios, results in fetal compression in the uterus. Fetal compression causes bony abnormalities of the limbs and face, deficiencies in intrauterine growth, and underdeveloped lungs. Obstruction can also result in body salt disturbances and renal failure. Patients may experience continuing difficulties even after the valves have been surgically opened (see later section). These persistent problems include partial renal failure, vesicoureteral reflux, and the inability of the bladder to void properly.

**What causes posterior urethral valves?**

Posterior urethral valves are found almost exclusively in boys. The incidence of PUV is somewhere between 1/5000 to 1/8000 males. The precise origin of PUV is not known. It is known that this condition is congenital, but not genetic. In other words, cases of PUV are sporadic and found at birth, but this condition is not passed from one generation to the next. Familial cases of PUV's are exceedingly rare.

**How is the diagnosis of PUV's made?**

A special x-ray of the urethra and bladder called a **voiding cystourethrogram** can show the obstruction of the posterior urethra and can also show vesicoureteral reflux (seen in 30-50% of patients). An ultrasound of the kidneys can also show the dilation of the ureter and kidney and give us an idea of the amount of renal tissue. In addition, special blood tests can be performed which assess the function of the kidney (creatinine - normal level in a term infant is 0.1 to 0.4 mg/dl). A good history and physical exam also plays an important role.

With widespread use of prenatal ultrasound, more PUV patients are being discovered prior to birth (antenatal diagnosis). Antenatal ultrasound may reveal dilated kidneys and ureters as well as oligohydramnios.
How are patients with PUV’s treated?

Initially, in infants with severe obstruction, a small catheter is placed into the bladder to relieve the obstruction. In the meantime, fluid and proper salts are replaced. If lung function is poor, mechanical ventilation is provided until he is able to breath on his own. If there is infection, antibiotics are given. Posterior urethral valves are classically treated by disrupting the valves with small knives or special hooks with exposed electrodes. These procedures are performed without a surgical incision, but rather directly through the urethra (endoscopic surgery).

If valve ablation does not improve renal function, as sometimes occurs in severe cases, other surgical options are necessary. These options include diverting the urine flow above the level of the urethra. Vesicostomy is opening a small portion of the bladder directly to the skin, while ureterostomy is the opening of a portion of the ureter directly to the skin. Sometimes a pyelostomy is performed whereby the urine from the kidney is brought directly out the skin overlying the kidney. These measures optimize the flow of urine from the kidneys and thus improve kidney function and minimize infections. The drawback to such operations is the need to perform further surgery in the future to bring the urine flow back down to the bladder. In certain patients, continued renal failure may require kidney transplantation. Occasionally, the bladder becomes a small capacity, high pressure system that may result in urinary incontinence, infections, and continued damage to the kidneys. Bladder augmentation (increasing the capacity of the bladder by using a piece of bowel as a patch on top of the bladder or cutting the wall of the bladder to allow its own inner lining to pouch out) may be necessary to correct these problems. In addition, bladder augmentation may provide a safe urinary reservoir for a future transplanted kidney.