Welcome to issue #10 of Bladdernews.

We hope you had a relaxing and enjoyable summer!

This issue’s feature article addresses “Hydronephrosis”, a condition that distends the kidney due to blockage of urine flow. You will learn about its symptoms and causes, and how it is treated.

Then, check out the pictures from the annual Spina Bifida Conference which took place June 30-July 3 in Orlando, FL. As sponsors of the conference’s “Kids Camp”, Astra Tech representatives had the opportunity to see first hand what sponsoring this event really means to the kids. Read one family’s special testimonial describing the positive impact that the Kids Camp had for their son! Also, don’t miss the personal stories from users just like you expressing how using a LoFric catheter has changed their quality of life.

Lastly, be sure to read the LoFric product updates and the handling tips for LoFric Primo and Hydro-Kit. There are also some back-to-school pointers to help parents minimize the stress associated with this time of year!

Enjoy this issue...happy reading!

**HYDRONEPHROSIS: Natural History, Causes, and Management**

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**DEFINITION**

Hydronephrosis (HN) means “dilated or swollen” upper urinary tract (kidney). The word “hydro” represents “water” and “nephro” represents “kidney”. It is not a true disease, but rather a descriptive term used to identify the amount of renal dilation from another disease or condition that may be present. HN can involve one or both kidneys. Normally urine flows out of the kidney with low pressure. If this flow pattern is disrupted by dilation of the kidney placing increasing pressure on the delicate internal structures of the central urine collecting system, damage can result through loss of function.

**DIAGNOSIS**

The diagnosis of HN is most often done through a renal ultrasonography (RUS). Ultrasound is not painful and simply requires the placement of a conductive medium (jelly) and a small transducer (camera) on the abdomen that captures images of the kidneys by using sound waves. A computed tomography scan (CT) or magnetic resonance imaging (MRI) can also be used. These studies are much more involved and expensive, but provide superior imaging quality and detail. Urine and blood tests such as creatinine, blood-urea-nitrogen (BUN), and carbon dioxide (CO2) are routinely done.

It is difficult to define HN physiologically as it may, or may not, cause symptoms. Diagnosing symptoms in young infants and children is very difficult. If symptoms occur they can include: back, flank (side of the body between the pelvis or hip and the last rib), waist or lower abdominal pain (including the urinary system), or nausea and vomiting. Urinary symptoms associated with dysuria (burning with urination), incontinence, hematuria (blood in urine), and febrile urinary tract or kidney infections are more rare.

Hydronephrosis should be graded accurately in order to make good clinical decisions concerning the management and follow-up. There are currently two methods to grade the degree of HN. The simple classification system of “mild, moderate, and severe” is less accurate. While not the gold standard of classifying HN, it is still used widely.

The Society for Fetal Urology (SFU) has developed a more accurate numerical grading system for HN. The criteria utilize numbers 0-4 to describe the amount of renal dilation seen. The grading criteria focuses on the degree of renal dilation of the intrarenal collecting system (calyces), blunting or distortion of the calyces, and the degree of paranchymal thinning (the outer rim of kidney tissue). Grade 0 is no dilation, 1 represents the least amount of dilation, and grade 4 represents the most severe degree of renal dilation. In addition, the size and degree of dilation involving the renal pelvis and ureters can be measured in cases with more severe obstruction.

There are currently other methods for grading HN being researched and developed. The causes of HN can originate congenitally (in the womb) or after birth. It is important to understand that HN does not always mean there is obstruction of the kidney, as there can be other causes of the dilation. With the incorporation of prenatal ultrasounds, ante-natal HN is currently the most common diagnosed prenatal problem.
Ureteral pelvic junction obstruction (UPJ) is due to imaging and no surgery. Imaging is significant, it can be followed with imaging and drainage is slow but clinically insignificant, it can be followed with imaging and no surgery.

**CAUSES**

1) **Uretero-vesical junction obstruction (UVJ)** is ureteral dilation seen in conjunction with HN, from an obstruction at the level of the bladder. Ureteral dilation can result from urine stasis and overstretching of the tissues referred to as hydroureteronephrosis or congenital megaureter (non-refluxing). UVJs can compromise renal function and may need surgical repair. Some forms of renal and ureteral dilation can result from urine stasis and overstretching of the tissues referred to as hydroureteronephrosis or congenital megaureter (non-refluxing). If renal function is not compromised and drainage is slow but clinically insignificant, it can be followed with imaging and no surgery.

2) **Ureteral pelvic junction obstruction (UPJ)** is an out-flow obstruction at the level of the kidney. Here the ureter has a narrowed, kinked, or weak area that does not peristalsis or move urine through efficiently and quickly to the bladder. Another cause of UPJ is outflow stasis of urine as the ureter passes over a crossing vessel (most commonly on the left side), that acts like a speed bump in the road that slows drainage. Sometimes a child may be born with a UPJ that is not identified at birth, and symptoms present intermittently and later in life. These are called intermittent UPJs and most often have to be diagnosed with an ultrasounds, CT scans, and associated symptoms (renal colic) during the symptomatic episode. If renal function and drainage is compromised, or symptoms persist, this condition needs surgical correction.

3) **Vesico-ureteral reflux (VUR)** is an abnormal “back wash” of urine from the bladder “back up” through the ureter and into the kidney. This condition is more common in girls than boys (4:1 ratio). VUR can be harmful to the kidneys during congenital development. After birth, VUR is generally not dangerous, unless there are associated episodes of febrile urinary tract infections or pyelonephritis (kidney infection). If renal function is compromised, or there is renal scarring from recurrent pyelonephrotic episodes, your doctor may recommend surgical correction.

4) **Posterior urethral valves (PUVs) or anterior urethral valves (AUVs)** is an out-flow obstruction along the urethra. The urethra is the small tube coming from the bladder where urine exits the tip of the penis. Small flaps of abnormal tissue can develop in the urethra acting like a dam or closed door. This condition only occurs in males and can cause dangerous back pressure (HN) and back-flow (VUR) of urine to the kidneys as well as myogenic bladder failure. With an antenatal ultrasound, the ensuing HN is seen early along with abnormal amniotic fluid levels. Valve disorders should always have immediate management and surgical correction at birth.

5) **Elevated bladder pressures** that can develop from various bladder conditions can cause HN and are often referred to as “hostile-unsafe” bladder environments. Examples include neurogenic bladders from spinal cord injuries, spina bifida, spinal cord tethering, spinal tumors or masses, Prune Belly Syndrome, and neurogenic non-neurogenic bladders such as Hinman-Allen Syndrome.

6) **Duplicated renal collecting systems** may show HN as well. There are two types, partial and complete duplications. With this condition the inner collecting system of the kidney develops into two parts instead of one, with two ureter tubes exiting the kidney. If the ureters attach together midway down, like a fork in the road, and then enter the bladder as a single ureter, it is a partial duplication. If the kidney has no HN, it is simply a variant of normal. However, if the ureters stay separated all the way down to the bladder, it is a complete duplication. With a complete type, generally the upper pole ureter will obstruct and the lower pole will reflux (VUR). If one ureter misses the bladder completely (ectopic) and enters the urethra or vagina, continual incontinence is always a presenting symptom. Complete duplications often need surgical correction.

7) **Ureterocele** can be a cause of HN. This is a congenital condition where there is an abnormal bulging of the ureter at its insertion point into the bladder. This bulge or bubble can cause obstruction of urine into the bladder. Renal-Bladder ultrasound is the most sensitive test for diagnosis. If clinically significant obstruction is caused, surgical incision is warranted. Ureterocele incision can often create VUR by disrupting the “one way valve” mechanism of the ureter. These may ultimately need surgical excision of ureterocele with ureteral reimplantation.
8) Kidney stones, or calculus, can cause obstruction of urine resulting in HN. If the stones are large enough, they can get lodged or caught in 3 locations listed below:
   a) if the stone forms in the kidney, as it tries to exit out into the ureter, it can get stuck resulting in obstruction and HN.
   b) as the stone tries to move down the ureter into the bladder it can cause obstruction and HN, especially as it reaches the UVJ where the ureter enters the bladder.
   c) as the stone tries to exit the bladder, obstruction can be caused in the urethra. Renal and bladder calculi can produce very severe symptoms of pain. If the stones are not able to pass spontaneously they may need surgical attention.

9) Multi cystic dysplastic kidney (MCDK) will show several round, well-defined, cystic structures within the kidney that often look just like severe HN. These abnormally developed kidneys are generally found early in pregnancy through antenatal ultrasounds. The true diagnosis cannot be made until the child is born and completes a series of tests including a post-natal ultrasound, voiding study (VCUG), and a nuclear renal scan (DTPA). The renal scan concludes the final diagnosis as it tells whether the kidney has any function. MCDKs have no function and will involute (shrink up) over time. These kidneys very rarely ever have to be removed. Only if they fail to involute, grow larger disrupting other organs, rupture, develop unmanageable high blood pressure, or if severe infection develops in the affected kidney would it need to be surgically removed.

10) Physiological or transient dilation is another cause of HN. This type of HN is generally very mild and can “wax and wane” over time. It is often seen in newborns, during the first few days of life, as they are still in a state of renal dehydration. In a majority of pediatric cases, these are followed every 3-6 months through the first year of life, and in the absence of any clinical symptoms, patients are discharged as a “variant of normal” with no further follow-up. Sometimes HN is found unexpectedly as patients go through other abdominal radiographic work-ups for gall bladder stones, kidney stones, appendicitis, and various sources of abdominal pain or trauma.

11) Other more rare causes are pelvic or abdominal tumors such as a pediatric Wilms’ Tumor. Injury, trauma, radiation therapy, abnormal scarring to the urinary system from previous surgery or accident, blood clots, enlarged uterus during pregnancy, enlarged prostate (BPH), and very severe constipation or fecal impaction.

TREATMENT
Treatment depends on the severity and cause of the hydronephrosis. The goal is to relieve the cause of obstruction or dilation. HN can also resolve spontaneously with no intervention. Newer surgical fetal intervention techniques utilize a vesicoamniotic shunt, placed in the fetus’s bladder to drain it, bypassing the obstruction in the urethra (PUVs). Urine is then allowed to drain into the amniotic space where the fetus is.

Hydronephrosis in children should be followed closely by a pediatric urologist who specializes in the complete management, all the associated causes, and surgical correction of this condition. HN cannot be prevented but early identification and treatment of the causes significantly reduces the risk of complications such as irreversible kidney damage.

Halverstadt Center of Excellence Pediatric Urology Clinic is located at the Children’s Hospital, University of Oklahoma Health Sciences Center in Oklahoma City.

Jake is the clinic manager and a full-time provider who sees all types of urological conditions, runs the dysfunctional elimination clinic, performs all of the post-operative and CIC teaching for the majority urinary reconstructive surgeries, as well as performs and interprets all the video-urodynamic testing.

Special thanks to his attending physicians:

Do you think you have a UTI?

1. Are there times when you feel a frequent urge to urinate?
2. Have you experienced a burning sensation when urinating?
3. Has your urine ever looked cloudy, discolored, or had a strong odor?

If you answered YES to any of these questions, you may be experiencing a urinary tract infection (UTI) and you are not alone. UTIs are the second most common type of infection. For women, 1 in 5 will be affected during their lifetime (statistics from the “National Kidney and Urologic Diseases Information Clearinghouse” website).

Whether you have experienced a UTI or not, it is a good idea to be aware of the symptoms and causes. This new pamphlet shows steps you can take to avoid them.