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THE TOP TEN
Pediatric Urological Diagnoses in Utah and the Great Basin

Why a booklet on Pediatric Urology?
For most people, the subject of abnormalities of the kidneys, ureters, bladder and genitalia is unknown territory! We have heard of adult problems, like prostate cancer and kidney stones, but most people know very little about the problems that infants and children are born with and the available treatments for their conditions.

What is Pediatric Urology?
Pediatric Urology is the surgical specialty that deals with problems of the genitalia and urinary tract of children. Most often, children are born with the problems that we see in Pediatric Urology. However, sometimes these problems are not known until children develop a urinary tract infection, such as a bladder or kidney infection. Sometimes problems with the kidneys are found on ultrasound before babies are born.

How common are the problems?
Most people are unaware of the common urological problems of children. This is because they are not often discussed outside the family. Most people have heard of urinary tract infections, but do not know that these infections are associated with anatomical abnormalities in approximately 30% of all children who have them, and even a higher percentage in boys. Among the genital defects, abnormalities of the penis are the most common followed by undescended testicles. In the operating room at a children’s hospital, urological cases are fourth in the total number of cases, following ENT (ear, nose and throat), general surgery, and orthopedics.

Who cares for Pediatric Urological Problems?
Pediatric urologists are surgical specialists who have trained in surgery and general urology after medical school and who then have completed fellowships specifically in pediatric urology. Pediatricians often manage medical problems of the urinary tract and genitalia. Nephrologists are specialists in medical diseases of the kidney, and endocrinologists specialize in endocrine problems affecting the kidneys, like diabetes, and of some problems of the genitalia, like ambiguous genitalia. Pediatric nurse practitioners work closely with patients whose problems require biofeedback and other behavioral or motor training.

Patients may be referred to urology after seeing a pediatrician or family practitioner for a general health evaluation. The primary care practitioner asks for a consultation.
In the year 2007, the top ten combined medical and surgical diagnoses overall were:

- Vesicoureteral Reflux
- Hypospadias
- Urinary Tract Infections
- Undescended Testes
- Bladder Dysfunction
- Abnormalities of the Foreskin
- Ureteropelvic Junction Obstruction
- Complications of Circumcision
- Neurogenic Bladder
- Nocturnal Enuresis (bedwetting)

When we separate out the patients most often seen by the nurse practitioners (Bladder Dysfunction, Bedwetting and Urinary Tract Infections), the most common problems seen by the Pediatric Urologists are:

- Hypospadias
- Vesicoureteral Reflux
- Undescended Testis
- Miscellaneous Disorders of the Penis
- Ureteropelvic Junction Obstruction
- Disorders of the Foreskin (Phimosis)
- Meatal Stenosis (abnormal tightness of the opening of the penis)
- Urinary Tract Infections
- Surgical Complications of Circumcision (abnormal scarring and adhesions)
- Hernias and Hydroceles

Other significant abnormalities of the genitalia and urinary tract are significantly more rare. For this booklet we will focus on the common abnormalities seen and questions most frequently asked.
Ultrasound: This is an easy test, similar to those obtained during pregnancy. A gel is applied to the abdomen and a wand (or transducer) is placed on the skin. Sound waves are transmitted through the skin to the inner organs (kidneys, bladder) and an reflected back to the transducer. An image is obtained that gives excellent information about the shape and size of the kidneys, ureters, and bladder.

Voiding Cystourethrogram (VCUG): This test is a little bit more invasive, because a catheter must be placed in the bladder. It is an important one, though, because some of the most common abnormalities of the urinary tract cannot be seen any other way. Children with vesicoureteral reflux may have a perfectly normal ultrasound, but have a very abnormal VCUG. During the test, a technician places a catheter in the bladder and fills the bladder with contrast material (dye). As the bladder is filling, the contrast shows the shape of the interior of the bladder, and sometimes the ureters. Then the patient voids, and we may see contrast going to the kidneys as well as out the urethra. In both boys and girls, this test gives valuable diagnostic information about the whole urinary tract. Additionally, the first image gives good information about the bowels - whether there is constipation or distension.
Nuclear Cystogram: This test is sometimes used for initial screening for vesicoureteral reflux, but more often used for follow-up. It is similar to a VCUG but rather than placing contrast in the bladder and taking x-rays, the technician places an isotope into the bladder and measures the counts during bladder filling and voiding. This test uses less radiation than actual x-rays but there is also less anatomical detail in the image. Therefore it is used less often as a first test.

Renal Scan: This test gives information about the function of the kidneys. It measures blood flow to the kidneys, uptake of the isotope, and excretion of urine. It gives diagnostic information about scars of the kidney, urinary obstruction or blockage, and comparison of function between kidneys. This is important information when a blockage is suspected based on ultrasound information, or when multiple episodes of kidney infection have resulted in scarring. The test involves placing an intravenous catheter (IV) for the isotope, and a bladder catheter to drain the urine. The isotope is run through the IV, and another medicine called furosimide (Lasix) is given at a specified time to increase the urine flow. The uptake and excretion of the isotope are then measured.
Computed Tomography (CT): This test is especially useful for visualizing kidney stones and kidney masses or tumors. Since these are relatively rare diagnoses in children, it is less frequently ordered.
Hypospadias and Disorders of the Penis

Who knew? Hypospadias, a disorder of the penis whereby the hole, or meatus, is abnormally located on the underside of the penis is the most common surgical abnormality of the genitalia. This abnormality of the penis was described by the Egyptians and Greeks.

In Utah, this abnormality affects close to 1:125 baby boys.

When the hole is in the wrong location, the foreskin is usually also abnormal, and does not cover the glans (or head) of the penis symmetrically. The underside of the foreskin may look like it has already been circumcised. In addition, the penis frequently curves downward, a condition known as "chordee".

Most cases of hypospadias are fairly minor and can be corrected in a single outpatient operation when babies are 6 months of age. More severe cases may require additional operations to correct. The operations take between one and three hours to do and must be done by specialists with expertise in reconstruction of the penis. The goal is to create a new urinary tube to bring the meatus or hole to the tip of the penis and to straighten the chordee. Often surgeons make use of the foreskin to create a new urinary tube or to cover the underside of the penis after correction of the chordee. There are many described operations for hypospadias, but in principle they break down into three variations: hinged or unhinged tubes, flaps, or grafts.

For the more complex operations, patients usually will require a urinary drainage tube for up to a week or two.
Complications of hypospadias repair include bleeding or hematoma, a fistula (or secondary hole that develops after surgery) recurrent curvature, or asymmetrical skin healing. Hematomas rarely require re-operation but most of the other complications do. The complication rate for hypospadias repair is between 5% and 20% depending on the complexity of the surgery and the age of the patient. Infants tend to do better than older children who are already toilet trained.

Other Congenital Abnormalities of the Penis:

- **Chordee:** Downward curvature of the penis
- **Torsion:** Rotation of the shaft or the shaft skin (usually to the left)
- **Webbing:** Webbed angle between the penis and scrotum
- **Buried or Hidden Penis:** Penis obscured by pubic fat
- **Phimosis:** Tight opening of the uncircumcised penis preventing retraction of the foreskin
- **Epispadias:** Opening of the urethra (urinary tube) at the top of the head of the penis.
- **Corporal Disproportion:** Asymmetrical development of the shaft of the penis.
- **Hooded Foreskin:** Asymmetrical development of the foreskin
- **Megameatus:** A variant of hypospadias, with an enlarged meatus.

Other abnormalities seen as a result of previous circumcision:

- **Meatal Stenosis:** Tight hole or meatus. This most commonly is diagnosed after toilet training when patients have a sideways or upward urinary stream. It is seen almost exclusively in patients circumcised at birth.
- **Adhesions:** Skin bridges or scarring along the circumcision scar line.
- **Phimosis:** Tightening of foreskin or shaft skin over the head of the penis after a circumcision due to "turtlenecking" of the skin and subsequent scarring.
Few issues in pediatrics and urology generate more emotion and discussion than circumcision. The pendulum swings back and forth regarding recommendations from the American Academy of Pediatrics and other professional and patient advocacy groups. The answer is that there is no right answer. Factors that go into a family's decision to circumcise a baby boy include religion and cultural norms and anatomy. For Jewish and Muslim boys, for example, there is a strong religious imperative for circumcision. For many families, if the father and or brothers are circumcised, the new baby will be circumcised as well.

In most European, Asian and Hispanic cultures, children are not circumcised. Complicating the decision is the fact that some insurance carriers and Medicaid do not cover elective circumcision. Never-the-less, there are some compelling arguments in favor of circumcision from the epidemiological point of view. Studies have shown that many sexually transmitted diseases, including HIV/AIDS are less common in and less commonly transmitted by circumcised men. Especially during the first year of life, there is a decreased risk of urinary tract infections in circumcised boys.

There can be complications of circumcision, such as meatal stenosis or adhesions (see above) and complications of no circumcision such as phimosis or infections under the foreskin (balanitis). In summary, each family or individual must make its own decision.
Urinary tract infections can affect girls and boys, men and women at any time in their lives. However, UTIs in babies and children often are related to abnormalities of the urinary tract that need special attention. This is why doctors order x-rays for children diagnosed with UTIs. Certain behaviors commonly associated with toilet training can also lead to UTIs. Constipation and holding the urine are some of the most common behaviors leading to UTIs. It is important to find out why children are getting UTIs in order to prevent more serious complications, such as kidney infections (pyelonephritis) which can lead to scarring of the kidneys.

Children are more prone to UTIs at certain ages. For boys at any age it is rare to have a UTI. However, during the first year of life, while in a diaper, and especially if uncircumcised, they are at greater risk. Most urologists recommend an ultrasound and a VCUG to make sure that the child does not have vesicoureteral reflux, which can be missed on an ultrasound alone.

Girls are more prone to infections both in the newborn period and during toilet training. Since girls are more likely to have reflux than boys, it is important to diagnose it as early as possible. In children with normal ultrasounds and x-rays, the reasons for UTIs usually include holding behaviors such as constipation and infrequent voiding. Some children will also avoid drinking fluids, which contributes to UTIs by minimizing washout of the bladder.

Finally, some people seem to be more prone to UTIs for unknown reasons. These may have to do with the constitution of the bladder lining, the immune system or other factors.
Vesicoureteral Reflux

In the normal condition, urine made by the kidneys travels in one direction down the ureters into the bladder where it is stored until urination. Some children are born with vesicoureteral reflux, wherein the urine "backwashes" up the ureter toward the kidney. By itself, reflux is not dangerous. However, if there are bacteria in the bladder, a simple urinary infection can turn into a kidney infection if bacteria are washed up to the kidney. Also, reflux can be an indication of high pressures in the bladder due either to true obstruction or to neurogenic bladder or dysfunctional voiding.

The average age at diagnosis is 2-3 years, during toilet training, and three quarters of the patients are girls. Reflux may run in families. As many as half of the children of mothers who had reflux will also have reflux. As many as one third of the siblings of refluxers will also have reflux.

Reflux occurs in about one in one hundred children, but is clustered in families of refluxers.

Diagnosis of Reflux: Reflux is suspected in any boys with UTIs or in newborn or young girls. A VCUG (voiding cystourethrogram) is an x-ray test where contrast or "dye" is instilled into the bladder through a catheter. If reflux is present, the contrast will be seen going up to the kidneys during the study. A nuclear cystogram is like the VCUG except that instead of using an x-ray machine to take pictures, a detector machine is used to measure the isotope in the bladder or ureters. (See photos on pages 3 and 4) It is good for screening or follow-up but doesn’t give the anatomical detail for diagnosis that the VCUG does. An ultrasound test is generally done at the same time to rule out any other anatomical defect of the kidneys.
Management and treatment of reflux: Successful treatment of reflux involves managing other issues that contribute to urinary tract infections and high pressures of the bladder. Chief among these is constipation, followed closely by dysfunctional voiding and neurogenic bladder. (see below) Even though surgery can correct reflux and many children will outgrow it, they may still have recurrent UTIs and pyelonephritis (kidney infections) if the behavioral component and the underlying bladder function is not addressed.

Medical management may include antibiotic prophylaxis, especially in babies at highest risk for kidney infections and subsequent scarring. Many people worry that chronic antibiotic therapy is unhealthy for children or the environment. This is indeed a concern, and it is a strong argument for managing the factors leading to reflux (see above). Additionally, some children are unable or unwilling to take medicine daily and it is an inconvenience for families.

Surgical options include endoscopic "STING" "Deflux" or "Hit" procedures, and open surgery with a standard incision. The endoscopic procedures depend on injecting a gel, dextranomer hyaluronic acid, near the ureter to provide resistance to backward flow of urine. It is 80-90% successful and can be repeated. Its advantage is that there is no incision and it is a fairly quick outpatient procedure. The disadvantage is that outcomes are not as good as standard (incisional) surgery.

**STING Procedure**
The standard surgeries, including the Cohen and Modified Lich procedures, involve repositioning the ureters with respect to the bladder. The ureter or ureters are repositioned to lie in a longer tunnel between the bladder lining and the bladder muscle wall. This provides a "flap valve" effect although no true valve is placed. The surgery may be done as an outpatient or as an overnight stay in the hospital. The success is 85% for the most severe reflux but almost 100% for milder grades.

**Cohen Procedure**

![Cohen Procedure Diagram]

**Modified Lich Procedure**

![Modified Lich Procedure Diagram]
Undescended Testis

Undescended testicles are testicles that have not come all the way down into the scrotal sac. Typically, they are diagnosed by the pediatrician or family doctor at well child checks within the first year of life. Most commonly, the undescended testicle is only on one side but occasionally can affect both. Urologists recommend that the testicles be brought down surgically within the first two years of life because studies have shown improved fertility in patients who have had the surgery done at this age compared to older children. Most children with undescended testes also have a hernia on the affected side which will also be repaired at the time of the operation.
Hernias and Hydroceles

Inguinal hernias and hydroceles are similar common problems of babies and children. They can affect boys and girls, but most commonly are seen in boys. They may also be hereditary. Hernia is the term used to describe a bulging at the groin that may extend into the scrotum, and usually has intestine or fatty tissue in it. A hydrocele only has fluid in it but may be similar in origin to a hernia.

**Hernias:** Hernias may be seen in newborn infants or they may develop in toddlers or children. Most children do not have the type of hernias that older adults do. They rarely involve a weakness of the body wall and do not require mesh for repair. Hernias in children most commonly are due to a failure of the processus vaginalis to close. The processus vaginalis is the continuation of the abdominal lining, or peritoneum into the scrotum. During development, when the testicle migrates into the scrotum from the abdominal cavity, it drags the abdominal lining into the scrotum with it. This channel usually seals itself, but sometimes does not. Sometimes it can be temporarily sealed, but open again when a child develops enough abdominal pressure to "pop" it open, around age two. Hernias do not heal themselves. They require an outpatient operation to close the open channel.

**Types of Hernias**

- **Normal**
- **Inguinal Hernia**
- **Complete Inguinal Hernia**

![Types of Hernias Diagram](image-url)
Hydroceles: A hydrocele is, literally, a "bag of water" in the scrotum. They most commonly occur around the testicle, but can also be found along the cord leading to the testicle. Newborn boys born with hydroceles often have the type that resolve over the first year of life. These are called "non-communicating" hydroceles, and they get smaller over time, rarely requiring surgery. Communicating hydroceles are much like hernias, because they "communicate" or connect with the abdominal cavity. They become larger and smaller as fluid flows into the abdomen or the scrotum. Normally, they do not heal themselves, but require surgery almost identical to surgery for inguinal hernias.

Ureteropelvic Junction Obstruction

Ureteropelvic Junction Obstruction (UPJ) or (UPJO) has been found more frequently since most mothers have ultrasounds during pregnancy. The problem is often identified at the 20 week ultrasound. It generally does not require early delivery. After the baby is born, an ultrasound is obtained. Depending on the degree of hydrenephrosis, another test may be done, a renal scan. If surgery is not immediately required, the doctor may recommend watchful waiting to see if the problem improves. More than fifty percent of the time, children will outgrow mild to moderate partial UPJO.
### Duplications of the Kidneys and Ureters

Duplications of the kidneys or ureters are common and may not be associated with any risk at all. On the other hand, sometimes duplications are associated with abnormal development of one or both ureters. The most common of these is when the upper half of the kidney does not fully develop and the ureter associated with it enters the bladder in an unusual (or ectopic) location. It can sometimes form "bubble" or ureterocele where it enters the bladder. Ureteroceles can obstruct the bladder neck and can also become infected because of poor drainage. They may need to be opened during early infancy, especially in girls, and will often need further surgery at a later time. A non-functioning upper half of a kidney may need to be removed, although, sometimes they may be left in place if the patient does not develop infections.

### Nocturnal Enuresis (Bedwetting)

Bedwetting is common in children. At age three, 60% of children are unable to sleep through the night without wetting. By age six, only 10% of children still wet the bed. Most children who wet the bed after age six have inherited this problem, and it is not uncommon to find one or several adult members of the family who also were bed wetters as children. If one parent had the problem, the chance is 45% that a child will also. If both parents wet the bed, the chance increases to about 75%. More boys have the problem than girls. Children do not willfully wet the bed, nor do they wet the bed as a result of emotional problems. However, if punished for wetting, they often will develop emotional problems. It is important to make sure that there are no other urinary problems, such as urinary tract infections complicating the picture. If so, these should be dealt with first.

Enuresis is described as "primary" when it has occurred since birth and the child has never had a dry spell. It accounts for 90% of all enuresis. Although most children are dry all day, some will also have daytime accidents. Children spontaneously out-grow primary enuresis at a rate of about 15% per year without medications or alarms. However, after age 7, it may become a social problem for some families.

Certain simple measures can be taken to "encourage" spontaneous resolution of enuresis. They include:

1. Encouraging your child to get up to urinate or waking him or her up when you go to bed.
2. Discouraging fluids after dinner.
3. Discouraging caffeine, including colas, teas, and chocolate in the evening.
4. Protect the bed with a plastic mattress cover rather than diapers or pull-ups.
5. Respond positively to dry nights
6. Respond gently to wet nights.

At age 8, if bedwetting persists, most children and parents are ready to be more aggressive. The next steps can be guided by a doctor, and include:

1. Bedwetting alarms. There are many of these available at toy stores and on the internet, most for about $50. They are small, lightweight, and sensitive to small amounts of urine.
2. Alarm clock. Some children respond well to these. Others sleep right through them. You will know your child best, but they are worth a try.
3. Medication. There are several medications available to help bedwetting.

Most children who are taking drugs for bedwetting also need to use an alarm. Your doctor will help you to decide the best option for medication, if indicated. The drawback to the medicine is that 50% of children go back to wetting when the medicine is stopped. However, they can be very helpful if the child will be traveling or going to camp, or in other situations where wetting may create significant emotional and social problems.
Resources for Parents and the Community

American Urological Association
www.auanet.org
www.urologyhealth.org/pediatric

European Society for Pediatric Urology
www.espu.org

Joint Committee of Pediatric Urology: A collaboration between the European Board of Paediatric Surgery and the European Board of Urology
www.espu.org/ej_index.php

Society for Pediatric Urology
www.spuonline.org

Constipation: An information page from the Children’s Hospital of Philadelphia
http://www.chop.edu/consumer/index.jsp (type “constipation” in Search box)

Hydronephrosis FAQs: Information page from the University of Chicago.
www.uchicagokidshospital.org/specialties/urology/patient-guides/hydronephrosis.html

Hypospadias: Information page from Cornell University
www.cornellurology.com/pediatrics/hypospadias.shtml

Hernias and Hydroceles: Informational page from Loma Linda University
www.llu.edu/lluch/pedsurg/inguinalhernia.html

Posterior Urethral Valves: Informational page from Stanford University
www.lpch.org/DiseaseHealthInfo/HealthLibrary/urology/puv.html

Spina Bifida: Informational page from University of California, San Francisco
www.ucsfhealth.org/childrens/medical_services/critical/sbifida/conditions/spina/signs.html

Ureteropelvic Junction Obstruction: Informational page from University of California, Los Angeles
www.uclaurology.com/conditions/Urete42.cfm

Vesicoureteral Reflux: Informational page from the University of Virginia Health System
www.Healthsystem.virginia.edu/uvahealth/peds_urology/vur.cfm