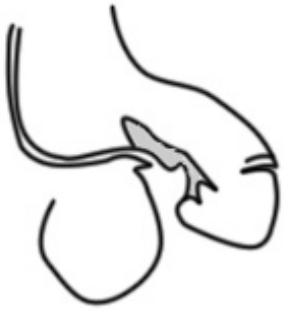
HYPOSPADIAS

SIDE VIEW

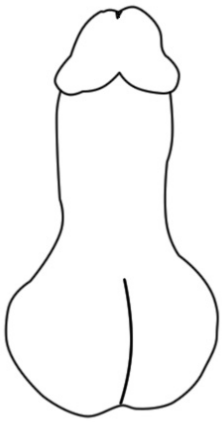
Normal

Hypospadia

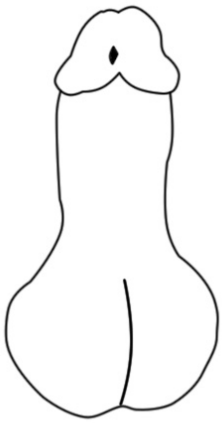


Severe Hypospadia with chordee

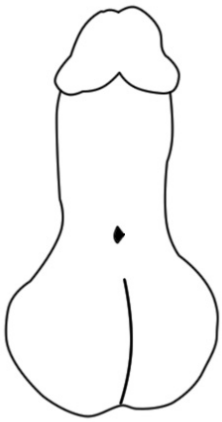
FROM BELOW



Normal



Hypospadia



Severe Hypospadia

What is hypospadias?

Hypospadias is a congenital (present at birth) condition affecting the penis. The penis contains a tube called the urethra which connects the bladder to the urethra opening (pee-hole) at the end of the penis. In hypospadias the pee-hole opens on the underside of the penis anywhere from just below the normal position (mild) to as far back as the base of the scrotum (severe). Sometimes a hooded appearance of the foreskin may be seen due to the underdevelopment of the foreskin on the underside of the penis. In more severe hypospadias the penis may be bent when stiff due to tightening of tissues (chordee) on the underside of the penis.

1 in 300 boys have hyposadias[1]. The exact cause is still unknown. However it is clear that hypospadias are associated with other conditions such as, undescended testicles or inguinal hernia[2]. All of which would be checked out by your doctor.

What problems hypospadias cause?

Hyposadias are not a life threatening condition and whilst in nappies the condition is relatively manageable[3]. The more severe the hypospadias, the more different from normal it will be to pass out urine. Urine will natural be directed downwards or backwards instead of forwards. Passing urine out at a urinal may be difficult. Later in life, if a bent penis is present, sexual intercourse may be uncomfortable and as a result psychological problems about being 'different' to normal are common[1].

How is it diagnosed and treated?

The goal of treatment is for urine to be passed in a forward way, for the penis to be straight when erect, and for the penis to look as normal as possible. Surgery before the age of 2 is carried out to create a new urethra and position the pee-hole at the top of the penis, and straighten out the penis. The operation is carried out under general anaesthetic and last anything from 1-3 hours[3].

Operation risks?

There is a small risk of bleeding and infection but these are minimized. For about 1 in 10 boys the original hole opens up again, meaning that your son may pass urine through two holes[3]. This can happen at any point after the operation, and will need to be fixed in

another operation. Occasionally, the new hole at the tip of the penis is too small, so your son will need another operation to make the hole larger.

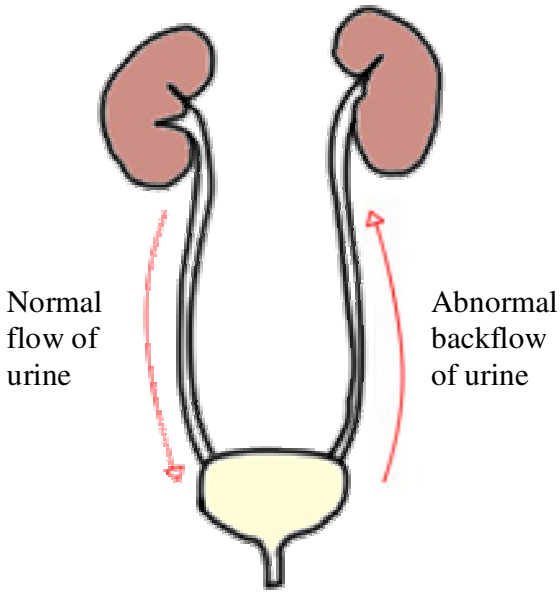
Home care

Your son will be sent home with a dressing and a catheter (small tube which passes through the penis to help with passing urine). It is common for the child to feel uncomfortable. Paracetamol (calpol) can be used to relieve pain. The dressing is usually kept on for a week. Antibiotics are given, to reduce the risk of infection. You should contact your GP if the dressing falls off, or if your child has a high temperature or if the wound is oozing[4].

VESICO-UTERIC REFLUX (VUR)

Background

The kidneys filter the blood and produce urine which flows down a tube (ureter) to the bladder. The bladder temporarily stores the urine until we pass the urine out. The ureter enters the bladder at angle, thus creating a flap like valve thus preventing the backflow of urine up the ureter towards the kidney.



What is vesico-ureteric reflux (VUR)?

VUR occurs when there is a problem with the ureter-bladder valve and so urine backflows from the bladder up the ureter towards the kidneys. In severe cases urine can reach the kidneys and cause damage.

VUR occurs 1 in 100 children and it is more common in the under fives and girls. It sometimes can run in families and some children can grow out of it[5].

What are the symptoms of VUR?

VUR is suspected in any child with repeated urinary tract infections (see UTI section for symptoms). In severe VUR the infection (bacteria) can reach the kidneys and cause damage and decline in kidney function.

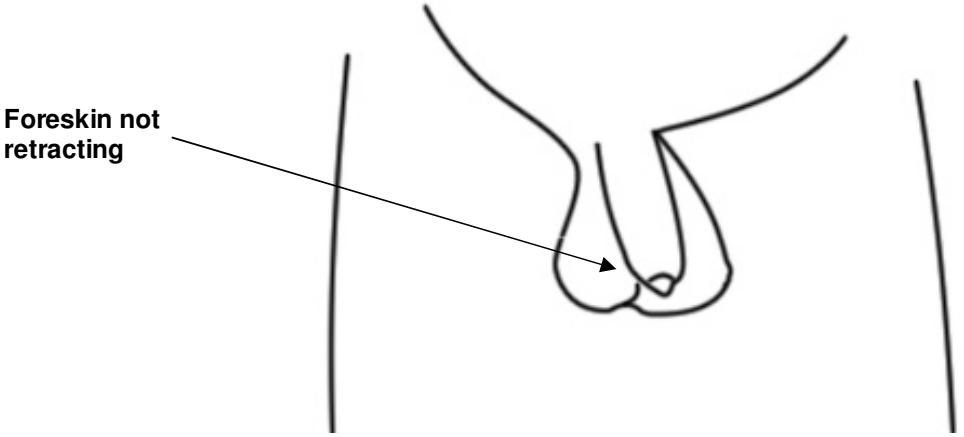
How is VUR diagnosed?

Ultrasound and VCUG (voiding cysto-urogram) are the best methods for detecting VUR. Ultrasound uses sound waves to create a picture of the structure of the kidneys, ureters and bladder; it is non-invasive procedure. VCUG determines the degree of reflux by viewing the flow of a contrast liquid which shows up on x-ray. The contrast liquid is inserted into the bladder through the urethra via a catheter. VCUG is slightly more invasive than ultrasound[6].

What is the treatment for VUR?

Treatment is conservative; the child is put on low dose long term antibiotics to prevent repeated infections. Hopefully this will allow the child enough time to grow out of the condition. Bearing in mind antibiotic resistance is on the increase; in some cases a procedure known as endoscopic sub-urethral injection can be carried out, where a safe compound is injected into the valves under the vision of a camera placed in the bladder through the urethra (pee-hole)[7]. Thus avoiding the need for open surgery. The choice of treatment is dependent upon the clinical case and in some cases surgery may be favourable. Surgery is offered to reimplant the ureters to create new bladder-ureter valves, thus preventing backflow.

PHIMOSIS



Phimosis is a condition where the foreskin is unable to retract over the head of the penis. All boys are born with the inability to retract their foreskin, but this does not amount to having phimosis as the foreskin in a newborn is tight and often it sticks. Separation occurs naturally and by the end of the first year only 50% have achieved this and by 3 years only 89%[8]. Majority would achieve separation by 16-17. True phimosis is defined as scarring of the tip of the foreskin and it is usually due to Balanitis Xerotica Obliterans which occurs in 0.4 cases/1000 boys per year[9].

What are the symptoms and how is it diagnosed?

Cleaning underneath the foreskin can be difficult; bacteria can accumulate and cause infection. Diagnosis is made by performing a routine physical examination.

What is the treatment for Phimosis?

Conservative treatment is opted, as in many cases the foreskin will retract as they boy grows. Parents are advised not to forcefully retract the foreskin as it may cause permanent damage. For true (pathological) phimosis circumcision is offered to completely remove the foreskin. The choice of treatment is based upon the clinical scenario, all of which should be discussed by your specialist.

URINARY TRACT INFECTION (UTI)

UTI occur when bacteria (usually from the bowel) get into the urine. Bacteria thrive in the urine; they quickly multiply and cause infection. The infection can affect any part of the urinary tract (i.e. kidneys, ureters, bladder, urethra) but mostly it causes infection in the bladder. Nearly 1 in 20 boys and more than 1 in 10 girls have at least one urine infection by the time they are 16 years old[10]. Children under five years are the most commonly affected.

What causes UTI?

Research has shown that any retention or stagnation of urine in any part of the urinary tract acts as an ideal breeding ground for bacteria. Various conditions cause retention some of the more common are[11]:

- **Constipation** – an impacted rectum can press on the bladder, thus preventing it from emptying completely.
- **Dysfunctional elimination syndrome** – this is when a child consciously holds onto the urine. There is no physical cause.
- **Structural abnormality** - e.g. VUR (see VUR section).

What are the symptoms of UTI?

UTI can present with many symptoms, in young children some more common symptoms are[12]:

- Fever (high temperature)
- Vomiting and/or diarrhoea
- Drowsiness
- Crying, going off feeds and generally unwell
- Appear to be in pain
- Blood in urine (uncommon)
- Cloudy or smelly urine

In older children, the child may complain of pain on passing urine, a burning sensation and increased urine frequency and maybe bed wetting. If the kidney becomes infected, the child may report tummy, back or side of tummy pain.

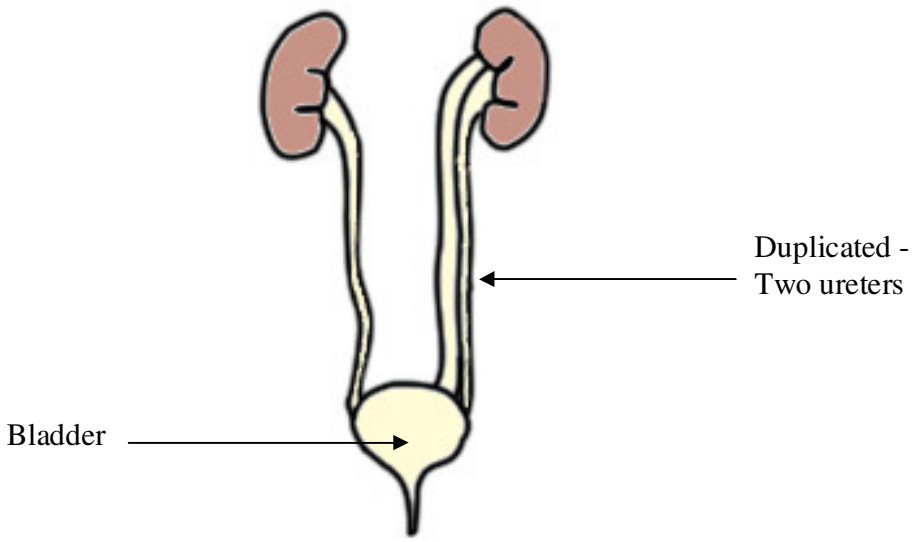
How is it diagnosed?

A simple test for the levels of bacteria on a sample of urine will confirm if the child has an UTI. The sample has to be clean as contaminated samples will give a false result.

What is the treatment?

A course of antibiotics usually clears up the infection and the child will recover fully with no concerns. To prevent further infections, it advised to drink plenty of water each day to wash out the urinary tract. Unresponsive/ persistent UTI raises suspicion of structural abnormalities and requires further investigations such as ultrasound and VCUG.

DUPLEX KIDNEYS



The urinary system consists of two kidneys one on each side, two ureters and one bladder. The kidneys filter the blood and create urine which passes through their corresponding ureters into the bladder. In Duplex kidneys sometimes either part or all of the kidney ureter system on one side is duplicated. So instead of having one kidney on each side, your child may have one on one side and two on the other. Duplex kidneys occur in about 1% of the population. Although a duplex kidney does not always cause problems, in some children it may not work well leading to a kidney infection and a risk of long-term damage. There may also be problems with the ureters as well, either not connecting with the bladder properly or being swollen; a condition known as an ureterocele[13].

How is diagnosed?

- **Ultrasound** uses sound waves to create a picture of the structure of the kidneys, ureters and bladder; it is non-invasive procedure and it might demonstrate the presence of two kidneys.

- **VCUG** determines the degree of reflux and thus presence of VUR (see VUR section) by viewing the flow of a contrast liquid which shows up on x-ray. The contrast liquid is inserted into the bladder through the urethra via a catheter. VCUG is slightly more invasive than ultrasound.

What is the treatment?

Surgery is the most effective treatment. An operation called a hemi-nephrectomy is performed to remove the part of the kidneys causing the problems whilst leaving the good part behind.

The operation is done under general anesthetic and it lasts between 2- $2^1/_2$ hours[7]. The operation can either be done by keyhole surgery (laparoscope) or traditionally as open surgery. Both methods are successful and their use is dependent upon the case.

Operation risks

There is a small risk of bleeding during surgery; the need for a blood transfusion for the procedure is rare. The risk of infection is minimized by prescribing antibiotics after the operation.

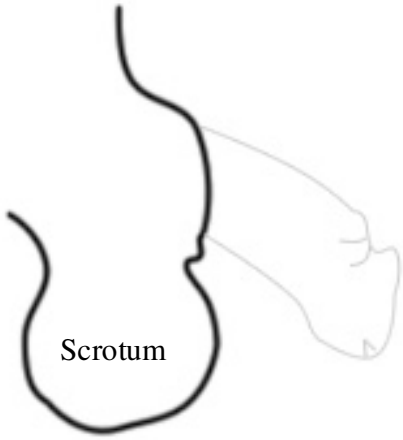
Home care

It is common for the child to feel uncomfortable after the operation. Paracetamol (calpol) can be used to relive pain. The dressing is usually kept on for a week. Antibiotics are given to reduce the risk of infection. Drinking plenty of water is advised to flush the urinary system in order to keep it clean. You should contact your GP if the dressing falls off, or your child has a high temperature or if the wound is oozing[4].

BURIED PENIS

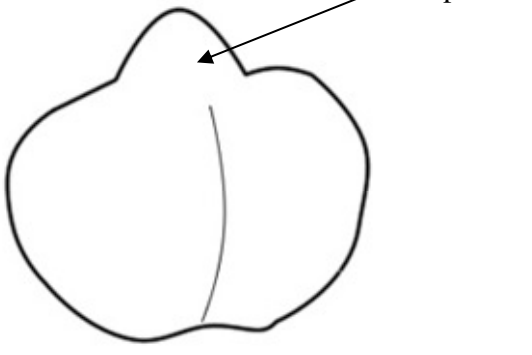
Buried (concealed) penis is a congenital disorder referred to a penis of a normal size which is hidden and not adequately exposed. The penis lacks the proper sheath of skin and lies hidden beneath the outer layer of the abdomen usually under a pad of fat.

SIDE VIEW



Buried
penis (faint outline shows expected position but in fact it is buried under the skin)

FROM BELOW



What causes buried penis?

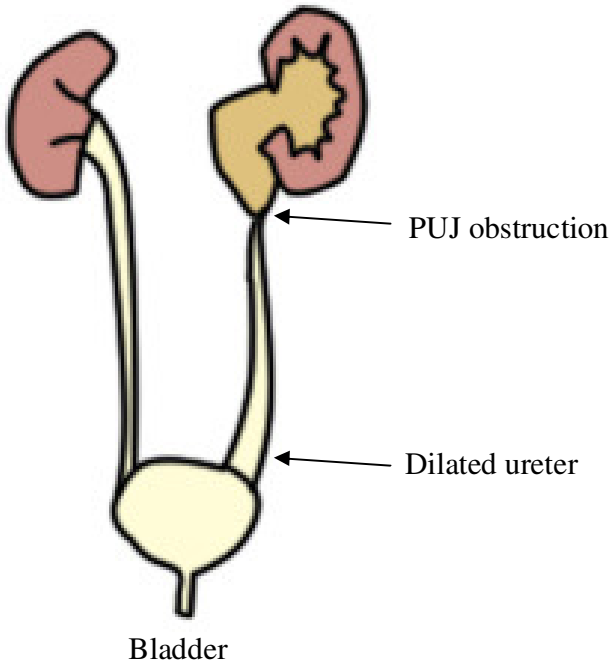
Some children are born this way (congenital) where a slight failure in penis development has restricted the full extension of the penis. Sometimes a prominent layer of pubic fat can conceal the penis, whilst for others it happens after an incorrect circumcision[14]. It is common in infants and toddlers, and occasionally seen in older children and obese adolescents.

How is buried penis diagnosed and treated?

Diagnosis is made by routine physical examination. If the penis can be easily exposed by gently pulling on it or by pressing down on the surrounding fat pad, then conservative management is an option, as this situation may correct itself over time. Sometimes surgery is needed for concealed penis. Either way results in full recovery with no concerns. Surgery is strongly recommended after the age 3 to aid voiding standing up during the toilet training period[15]. The choice of treatment is dependent upon the clinical scenario and specialist medical help should be sought.

PELVIC-UTERIC JUNCTION OBSCTRUCTION (PUJ)

This is a narrowing at the junction between the kidney and ureter; the tube which carries the urine from the kidney to the bladder. This narrowing results in the blockage and subsequent build up of urine within the kidney[16].



What causes it?

In children the condition commonly presents at birth due to an abnormality in the structure at the ureter kidney junction. The cause of this abnormality is unknown. In adults the condition is usually secondary to kidney stones or UTI.

What are the symptoms?

Pain on the side of the abdomen or back is common especially after drinking large quantities of water. The impeded flow of urine at the ureter kidney junction causes urine to accumulate in the kidney. Pressure created by the urine causes the kidneys to enlarge (hydronephrosis) and over a long period of time the kidneys may become damaged. Damaged kidneys may present with blood in the urine (haematuria) and a decline in kidney function.

How is PUJ diagnosed?

Occasionally an enlarged kidney may be palpated during routine examination, but a diagnosis of PUJ is only confirmed by one of the following tests[17]:

- **Ultrasound** might show an enlarged kidney (hydronephrosis)
- **Intravenous urogram** (IVU). An IVU is a special x-ray test in which multiple pictures are taken of the kidney after a dye (contrast liquid) is given through a vein. The contrast demonstrates the narrow region of the PUJ, but the IVU does not always demonstrate PUJ obstruction.
- **Diuretic renogram** This is a more detailed kidney scan that on most occasions will demonstrate PUJ obstruction. For these, an injection of a chemical is given into the blood and pictures taken of the kidney. There are two main types, either MAG3 or DTPA.

What is the treatment for PUJ?

Initially for the first 18 months of life, a period of regular review is recommended to evaluate the extent of the obstruction. In some cases the obstruction may heal on its own. Persistent obstruction will require an operation called pyeloplasty to remove the damaged ureter section and reattach the ureter back onto the kidney. The operation can be done laparoscopically or traditionally via open surgery and it is very successful.

Operation risks

There is a risk that urine could leak from the join in the ureter. Placing a tube (blue stent) in the ureter to drain urine can help prevent this from happening. There is a small chance that the join may narrow so the flow of urine becomes blocked again, which may mean your child would need to have a second operation.

Home care

It is common for the child to feel uncomfortable after the operation. Paracetamol (calpol) can be used to relive pain. The dressing is usually kept on for a week. Antibiotics are given to reduce the risk of infection. Drinking plenty of water is advised to flush the urinary system in order to keep it clean. You should contact your GP if your child has a high temperature or if the wound is oozing[4].

INCONTINENCE

The bladder is a storage vessel for urine before it is passed out. Controlling this activity involves nerves, muscles, the spinal cord, and the brain. At birth your child has no control over their bladder and so the child involuntary passes urine out. Over time the system matures and eventually the child will gain conscious control of the bladder and learn to void when it is necessary and appropriate[18]. Failure in the control of the system leads to involuntary passing of urine which is known as incontinence. Incontinence is classified as either organic or functional.

What is organic Incontinence?

Organic incontinence is due to an underlying disease process, which can either be neurogenic (nervous system) or structural. Neurogenic forms include sacral agenesis, cerebral palsy, spinal cord injury, and tethered spinal cord. Structural incontinence refers to developmental abnormalities of the lower urinary tract that interfere with the urinary system's ability to hold, store, or evacuate urine.

What is functional incontinence?

This is the presence of incontinence where no structural or neurological abnormality can be found.

How common is incontinence?

Incontinence happens less often after age 5: About 10 percent of 5-year-olds, 5 percent of 10-year-olds, and 1 percent of 18-year-olds experience episodes of incontinence. It is twice as common in boys as in girls[19].

How is it diagnosed?

Diagnosis is made on an accurate voiding history and physical examination screening for any neurological signs. Further tests include screening the urine for UTI and ultrasound. Slightly more invasive is VCUG (voiding cysto-urogram) which allows for the flow of urine to be viewed by tracking a contrast liquid which shows up on x-ray. The contrast liquid is inserted into the bladder through the urethra via a catheter.

What is the treatment?

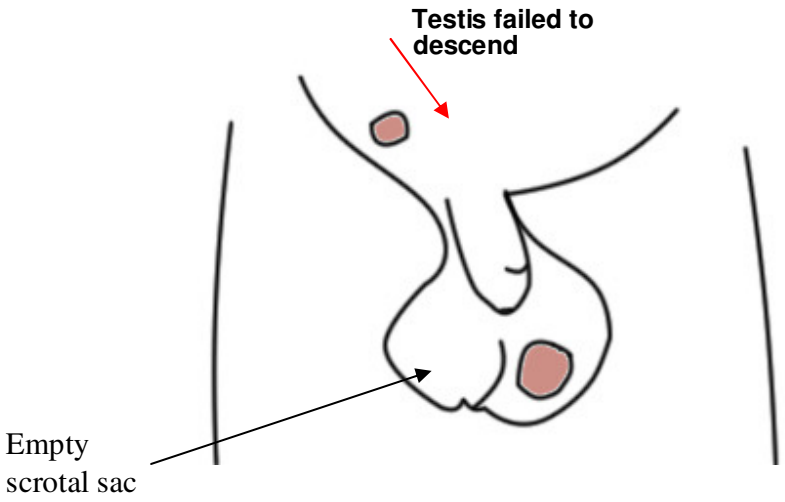
Many children improve over time, and some are symptom free. Depending upon the cause of the incontinence any of the following treatments can be used[19]:

- **Conservative** – Many children overcome incontinence naturally without treatment as they grow older. The number of cases of incontinence goes down by 15 percent for each year after the age of 5.
- **Medication** – Some children respond to medication which facilitates the production of a hormone called ADH (anti diuretic hormone) which is responsible for controlling the production of urine.
- **Surgery** – structural abnormalities can be removed in order to alleviate symptoms.

UNDESCENDED TESTICLES

In boys the testicles begin developing in the abdomen of the child during pregnancy. Near to the time of birth the testicles descend through a canal from the abdomen to the scrotal sac. At birth both testicles can be felt in the scrotal sac. Sometimes either one or both testicles cannot be felt, as they have failed to descend from the abdomen. This is known as undescended testicles.

Around 1 in 20 boys are born with undescended testicles and about 1 in 70 boys the testicles remain undescended[20].



What are the symptoms?

Apart from the missing presence of the testicles form the scrotal sac there are usually no symptoms. It is very uncommon for the child to be in pain unless the testicles become twisted, which is extremely painful and requires immediate help. The risk of testicle torsion is high due to the testicles not being in the right place.

Although there is no immediate risk; long term there is a high risk for the following[21]:

- **Cosmetic reasons**. Most boys would prefer to have two testes that look normal when they are grown up.
- **Fertility**. If the testes are left in the abdomen, there is a high chance that they will become infertile during adulthood.
- **Cancer**. The risk of developing cancer of the testes in adulthood is higher if the testes are left in the abdomen.

How is it diagnosed?

Routine examination of the scrotum and abdomen will determine the presence of testicles. Sometimes cold weather can cause the testicles to ride up the canal and present with a false impression of undescended testicles.

How is it treated?

Sometimes the testicles can descend on their own and so a period of watch and review for first 6 months is recommended. After 6 months an operation called an orchidopexy is recommended. This is a short operation where the testicles are brought down and placed in the scrotal sac.

Operation risks

The small risk of anesthesia and bleeding is uncommon due to the expertise of the surgeon and anesthetist. Occasionally there will be bruising in the groin area after the operation but this is expected. There is a small risk of the testicles becoming damaged during the operation; however the operation is very successful.

MULTICYSTIC DYSPLASTIC KIDNEY DISEASE (MCKD)

This is a condition present at birth where the kidneys have failed to develop in the child due to cysts. Cysts are little fluid filled sacs which occupy the kidney and prevent the kidney structure to form and thus carry out its specific function. Typically this condition only affects one kidney and it occurs in 1 in 4300 live births[22]. The cause of this condition is still unknown.

Are there any other abnormalities?

Typically MCKD is an isolated problem affecting one of the kidneys. MCKD in both kidneys is not compatible with life. However there is a risk of an associated abnormality which can be high as 51%. Such conditions include such as vesico-ureteral reflux, uretero-pelvic junction obstruction, or uretero-vesical junction obstruction (see relevant sections for more information).

How is it diagnosed?

In MCKD the kidneys are enlarged and so MCKD can be suspected before the baby is born in the womb by ultrasound. Further testing after birth can be carried out to ensure the accuracy of the initial diagnosis.

What is the treatment for MCKD?

Assuming that only one kidney is affected (life is not viable with MCKD in both kidneys), the child is unlikely to encounter any problems during pregnancy or life. One kidney is fully capable of supporting the body. Therefore initial management is to watch and wait to allow time to be certain of the diagnosis and severity of the condition. Sometimes the affected kidney will regress and disappear. However if it is felt necessary the affected kidney can be removed in a procedure known as a nephrectomy which may be performed by keyhole surgery; a less invasive method than open surgery.

Word count 3,609

[1] http://www.patient.co.uk/showdoc/23069152/ accessed 1/2/09
[2] Weidner IS, Moller H, Jensen TK, Skakkebaek NE. Risk factors for cryptorchidism and hypospadias. J Urol 1999;161(5):1606-1609. http://www.ncbi.nlm.nih.gov/pubmed/10210427?ordinalpos=2&itool=EntrezSystem2.PEntrez.Pubmed.Pubmed_ResultsPanel.Pubmed_RVDocSum
[3] http://www.ich.ucl.ac.uk/gosh_families/information_sheets/hypospadias/hypospadias_families.html accessed 27/01/09
[4] http://www.childrensurology.co.uk/HypospadiasRepair.htm accessed 27/01/09
[5] Sargent MA (September 2000). "What is the normal prevalence of vesicoureteral reflux?". *Pediatr Radiol.* 30 (9): 587–93. PMID 11009294
[6] Hollowell JG, Greenfield SP. Screening siblings for vesicoureteral reflux. J Urol 2002;168:2138-2141. http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=pubmed&dopt=Abstract&list_uids=12394743&query_hl=16&itool=pubmed_docsum
[7] Tekgul S, Riedmiller H, Gerharz E, Hoebeke P, Kocvara R, Nijman R, Radmayr C, Stein R. Guidelines on paediatric urology. Arnhem, The Netherlands: European Association of Urology, European Society for Paediatric Urology; 2008 Mar. p. 47-52
[8] Gairdner D. The fate of the foreskin: a study of circumcision. Br Med J 1949;2(4642):1433-1437. http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=pubmed&dopt=Abstract&list_uids=15408299&query_hl=9&itool=pubmed_docsum
[9] Oster J. Further fate of the foreskin. Incidence of preputial adhesions, phimosis, and smegma among Danish schoolboys. Arch Dis Child 1968;43(288):200-203. http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=pubmed&dopt=Abstract&list_uids=5689532&query_hl=11&itool=pubmed_docsum
[10] Mori R, Lakhanpaul M, Verrier-Jones K; Diagnosis and management of urinary tract infection in children: summary of NICE guidance. BMJ. 2007 Aug 25;335(7616):395-7.
[11] Urinary tract infection in children: diagnosis, treatment and long-term management, NICE Clinical Guideline (2007)
[12] http://www.patient.co.uk/showdoc/23068848/ accessed 22/02/09
[13] Coplen DE, Duckett JW. The modern approach to ureteroceles. *J Urol.* Jan 1995;153(1):166-177. [Medline].
[14] Maizels M, Zaontz M, Donovan J. Surgical correction of the buried penis: description of a classification system and a technique to correct the disorder. *J Urol.* Jul 1986;136(1 Pt 2):268-71. [Medline].
[15] Cromie WJ, Ritchey ML, Smith RC, Zagaja GP. Anatomical alignment for the correction of buried penis. *J Urol.* Oct 1998;160(4):1482-4. [Medline].
[16] Lebowitz RL, Griscom NT. Neonatal hydronephrosis: 146 cases. Radiol Clin North Am 1977;15(1):49-59. http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=pubmed&dopt=Abstract&list_uids=139634&query_hl=44&itool=pubmed_DocSum
[17] Grignon A, Filiatrault D, Homsy Y, Robitaille P, Filion R, Boutin H, Leblond R. Ureteropelvic junction stenosis: antenatal ultrasonographic diagnosis, postnatal investigation, and follow-up. Radiology 1986;160(3):649-651.http://www.ncbi.nlm.nih.gov/entrez/query.fcgi?cmd=Retrieve&db=pubmed&dopt=Abstract&list_uids=3526403&query_hl=27&itool=pubmed_docsum
[18] Goellner MH, Ziegler EE, Fomon SJ. Urination during the first three years of life. Nephron 1981;28:174-8.

[19] Jones EA. Urinary Incontinence in children. Chapter 12. http://kidney.niddk.nih.gov/statistics/uda/Urinary_Incontinence_in_Children-Chapter12.pdfAccessed 2/2/09
[20] http://www.ich.ucl.ac.uk/gosh_families/information_sheets/undescended_testicles/undescended_testicles_families.html
[21] Davenport M; ABC of general paediatric surgery. Inguinal hernia, hydrocele, and the undescended testis. BMJ. 1996 Mar 2;312(7030):564-7.
[22] http://www.childrenshospital.org/az/Site1318/mainpageS1318P0.html. Accessed 22/02/09