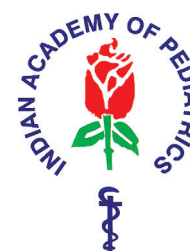


Indian Academy of Pediatrics (IAP)



GUIDELINES FOR PARENTS

Congenital Anomalies of Kidney and Urinary Tract



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11 FAQs on CONGENITAL ANOMALIES OF KIDNEY AND URINARY TRACT

Hydronephrosis

1. I am 5 months pregnant woman and my recent ultrasound shows that the baby has some swelling called "hydronephrosis" in both the kidneys. I am very worried whether my child will be normal, and should I continue with pregnancy or not?
2. I have recently delivered a baby who is 1-month-old and has a swelling in one of the kidneys. Ultrasound report says "hydronephrosis". How serious is it? Do we need any further tests?

Dribbling and Weak Urinary Stream of Urine

3. Doctor, we have noticed that my son, who is 3 months old, has a poor urinary stream and often cries during voiding? What could be the problems and does he need any test?

Cystic Kidney Disease: Autosomal Recessive Polycystic Kidney Disease/Simple Cyst

4. Doctor, my daughter who is 6 years old was having abdomen pain, so a general practitioner (GP) advised an ultrasonography (USG) scan which is

showing a cyst in the upper pole of right kidney (2 × 2 cm). What should I do next? Is genetic testing required?

5. Doctor, my husband has cysts in his kidneys, and my father-in-law also had some kidney problem? We are planning for our first child; what precaution should we take?

Phimosis/Hypospadias

6. We have noticed that the foreskin of my 15-month-old boy's penis cannot be retracted. Should we apply gentle force to pull it back?
7. My 4-month-old baby boy is passing urine through an abnormal hole on the inner side of his penis so the urine falls on his thighs and legs. What should be done?

Drugs Exposure to Mother in Antenatal Period Affecting Fetal Kidney

8. What painkillers can be given to a pregnant woman with severe backache? Can we use angiotensin-converting-enzyme (ACE) inhibitors for blood pressure control during pregnancy?

Single Kidney

- 9A. Doctor, should I be worried about my child having single kidney?
- 9B. Why did my child develop this problem?
- 9C. What steps do we have to take after birth to ensure that my baby stays well?
- 9D. As she/he has only one kidney; after birth and as my child grows up do we need to restrict any activities?

Multicystic Dysplastic Kidneys

- 10A. My 6-month-old son was found to have palpable right flank lump. On further evaluation, the doctor has stated that he has a nonfunctioning multicystic dysplastic right kidney (MCDK). What is MCDK and why did it occur in my child?
- 10B. Will my child need surgery for multicystic dysplastic kidney (MCDK)?
- 10C. How long do we need to keep my child under medical follow-up and what tests he will need regularly?

Horse-shoe Kidneys

11. Doctor, my child has been diagnosed to have "horse-shoe" kidney. What does this mean and what needs to be done?

Under the Auspices of the IAP Action Plan 2021–2022

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Congenital Anomalies of Kidney and Urinary Tract

Hydronephrosis

Q1

I am 5 months pregnant woman and my recent ultrasound shows that the baby has some swelling called “hydronephrosis” in both the kidneys. I am very worried whether my child will be normal, and should I continue with pregnancy or not?

Hydronephrosis is a medical condition which often results from an obstruction to the flow of urine from the kidney into the ureter (the tube which connects the kidneys to the urinary bladder). The obstruction can be secondary to narrowing in the urinary passage particularly happening at junction of the tube (ureter) and the pelvis or the junction of ureter and bladder. In addition, boys can have a membrane in their urinary passage in urethra which can also cause obstruction (posterior urethral valve—discussed later). Apart from obstruction hydronephrosis can also result from backflow of urine from bladder back to kidney (vesicoureteric reflux). Obstruction to the passage of urine either through narrowing or through vesicoureteric reflux result in accumulation of urine in central part of the kidney called

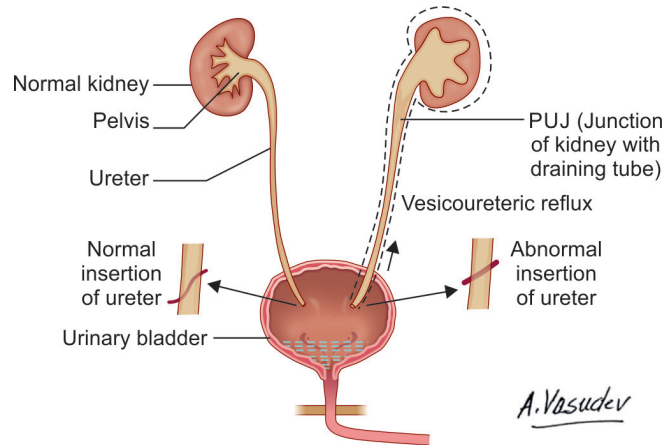


Fig. 1: Right side is the normal kidney and ureter/left side is abnormal kidney (hydronephrosis) and ureter (reflux). (PUJ: pelvic ureteric junction)

the pelvis, which is defined as hydronephrosis (**Fig. 1**). A bigger size of pelvis will need frequent assessment with ultrasound. Apart from this, we must also assess the other organs of the baby such as the heart, alimentary canal, brain and nerves, bones, etc. for any major defect and for the amount of amniotic fluid (liquor amnii), i.e., fluid in the womb. A decrease in the amount of amniotic fluid is due to the poor work of kidneys and needs repeated assessments by ultrasound. Decrease in amniotic fluid and/or involvement of both kidneys are considered as high-risk factors and need regular ultrasound assessment *in utero* as well as early ultrasound postdelivery. Usually, intervention is planned postdelivery and type of intervention depends on the etiology of hydronephrosis, i.e., narrowing of the urinary passage, membrane flap in the urethra in boys or reverse flow of urine from bladder to kidney. Termination of pregnancy is not recommended unless there are life-threatening abnormalities of other organs or a severe decrease in amniotic fluid. Fetal interventions, if required, are available at specialized centers.

Q2

I have recently delivered a baby who is 1-month-old and has a swelling in one of the kidneys. Ultrasound report says “hydronephrosis”. How serious is it? Do we need any further tests?

Mostly the hydronephrosis is detected on ultrasound done during the pregnancy. In 50% of these, the swelling subsides by the end of 3rd trimester of pregnancy. An ultrasound is done in these babies 1 week after the birth. If the size of renal pelvis (middle part of the kidney) is 10 mm or more in anteroposterior diameter (APD), a repeat ultrasound is done at 1 month of age. At 1 month of age, if the APD is persisting to be 10 mm or more the child will require further tests to find out the cause of persistent swelling. At a first step, the child will have a special X-ray test called as micturating cystourethrography (MCUG), which will rule out if the urine from the bladder is moving in the reverse flow to the kidneys (vesicoureteric reflux, **Fig. 1**). In case of boys, MCUG may need to be done earlier, if there are suspicions of posterior urethral valve (membrane in the urethral passage) such as significant bilateral dilatation of urinary passage or poor urinary stream/dribbling of urine. Till MCUG is done and vesicoureteric reflux is ruled out, children are kept on a small dose of antibiotic to prevent infection in the dilated urinary tract. A swelling <10 mm size does not need detailed testing although at least one more repeat ultrasound scan is usually advised. Once vesicoureteric reflux is ruled out, these babies are followed up with ultrasound at 3, 6, and 12 months of age and later till the swelling resolves. If the renal pelvis APD increases significantly (≥ 15 mm), then a nuclear scan (DTPA renal scan) may be required to rule out any obstruction to the outflow of urine from the kidneys (pelviureteric junction obstruction). A dilatation of >20–30 mm has a high chance of need for surgery. Children with significant hydronephrosis usually need a long-term follow-up and should be under joint care of pediatric nephrologist and a pediatric surgeon.

Dribbling and Weak Urinary Stream of Urine

Q3

Doctor, we have noticed that my son, who is 3 months old, has a poor urinary stream and often cries during voiding? What could be the problems and does he need any test?

The child needs evaluation as he may have developmental abnormality of the genitourinary tract like a membrane in the urinary passage [posterior urethral valves (PUV)] or narrowing of urinary passage (meatal stenosis). He will require blood pressure measurement, urine examination, kidney function tests, and ultrasonography now. Depending upon the results, we will be able to describe you the exact nature of the illness.

The most concerning abnormality which we are worried of is posterior urethral valve, i.e., PUV (membrane in the urethral passage) (**Fig. 2**), which can cause high pressure in both the kidneys leading to bladder damage as well as dilatation of kidneys and ureters. Confirmation of PUV requires a radiological test in which a catheter is passed in the bladder through which dye is instilled and X-ray films are taken. A pediatric surgical consultation is also required, since if confirmed, a surgical resection of the membrane will be required.

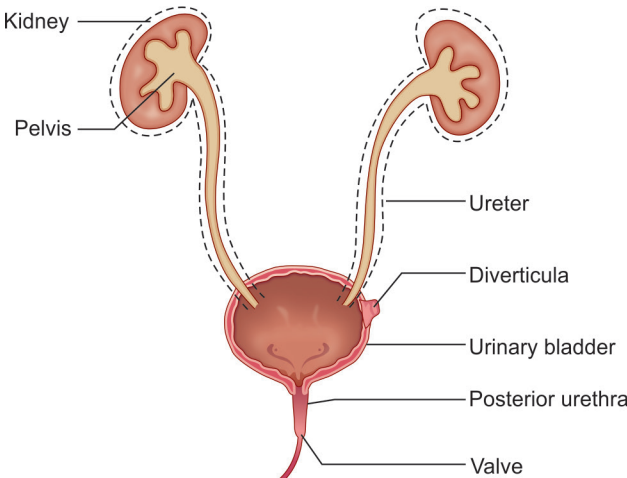


Fig. 2: Posterior urethral valve resulting in bilateral dilatation of kidneys and ureters (hydronephroureterosis) as well as thick bladder wall and outpouching of bladder wall (diverticula).

Cystic Kidney Disease: Autosomal Recessive Polycystic Kidney Disease/Simple Cyst

Q4

Doctor, my daughter who is 6 years old was having abdomen pain, so a general practitioner (GP) advised an ultrasonography (USG) scan which is showing a cyst in the upper pole of right kidney (2 × 2 cm). What should I do next? Is genetic testing required?

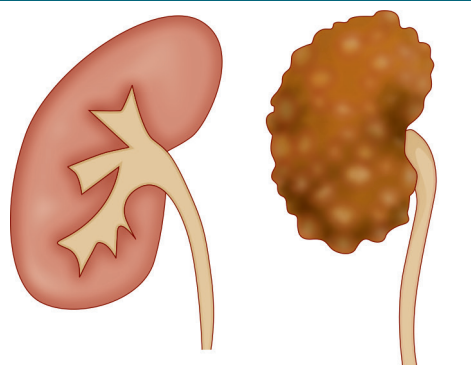
The ultrasound report mentions a single cyst (fluid-filled sac) of 2 × 2 cm, sharply defined with well-transmitted sound waves and absence of any echoes, this is likely to be simple cyst. Simple cyst has a prevalence of 7–10% in the general population, and usually does not cause significant problem. Your child will require annual follow-up only. Genetic testing is advised only if parents have history of cysts in their kidneys or the child in future develops multiple cysts. You need to be aware of this condition and mention to your local doctor, if your child develops persistent/recurrent fever or proven urinary tract infection.

Congenital Anomalies of Kidney and Urinary Tract

Q5

Doctor, my husband has cysts in his kidneys, and my father-in-law also had some kidney problem? We are planning for our first child; what precaution should we take?

The history indicates possibility of polycystic kidney disease (**Fig. 3**) in your family. Your husband will require some tests and nephrology consultation to characterize his condition so that we can know if there are chances of any recurrence in your baby. These are usually inherited/familial disease and with available genetic tests in many instances, it is possible to arrive at a definitive diagnosis.



Normal kidney Polycystic kidney
Fig. 3: Polycystic kidney disease.

Phimosis/Hypospadias

Q6

We have noticed that the foreskin of my 15-month-old son's penis cannot be retracted. Should we apply gentle force to pull it back?

The glans penis of a newborn baby is covered with foreskin (prepuce). Normally, the foreskin is not retractable due to adhesions which bind it to the glans penis. This *tight prepuce* is also called "physiological" phimosis and is commonly seen up to 3 years of age. *Paraphimosis* is where the foreskin cannot be returned to its original position after being retracted. Paraphimosis causes the glans to be painful and swollen and requires emergency treatment by a trained professional. Ballooning of foreskin occurs during urination and this helps in its physiological separation from the glans over time. Although it is possible to retract the foreskin in 40% of boys by 1 year of age, *parents should not attempt retraction at home* and wash only the outer surface of the penis during a bath. At 15–18 months pediatrician checkup, if retraction of the foreskin is possible, then parents can rinse the glans and inner foreskin with warm water regularly during bath. *True phimosis* is uncommon and usually occurs due to recurrent infections (balanoposthitis).

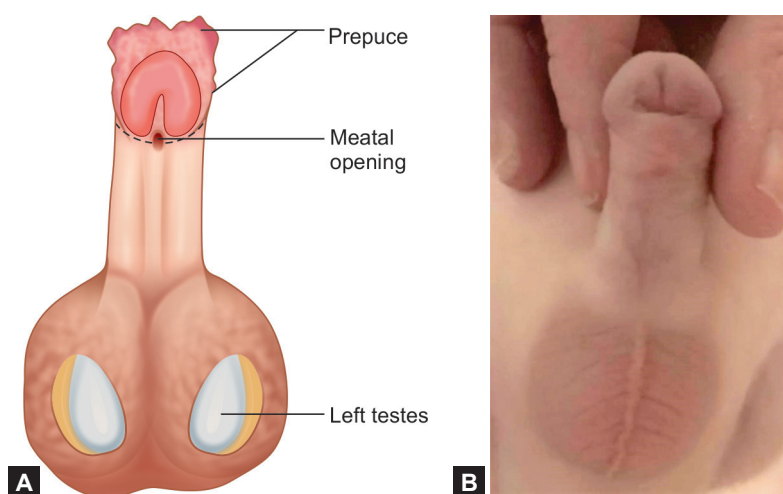
Treatment: Phimosis in children does not require any treatment. If not retractable by 2 years of age, local application of 0.5% betamethasone once or twice daily for 2 months is effective in 90–95% of children. If topical steroid application is ineffective, circumcision may be required. It is a surgical procedure which removes part or whole foreskin. It is safe and has been performed over many years. Treatment for paraphimosis is by pushing the foreskin ahead by application of a local anesthetic. Surgical correction by a slit in the foreskin or circumcision may be necessary in severe cases.

Q7

My 4-month-old son is passing urine through an abnormal hole on the inner side of his penis so the urine falls on his thighs and legs. What should be done?

Your child may be having “*hypospadias*”. This is a condition where the urinary opening is not in the correct place but can be anywhere along the underside of the shaft or even at the root of the penis near the testicles. Most commonly the opening is in the midline on the inner side, at the junction of glans and shaft of penis (**Fig. 4A**). The penis may bend at the end when erect. Part of the foreskin may be absent on the underside of the penis, so it looks like a hood (**Fig. 4B**). Hence, the stream of urine is directed downward toward the feet, legs, or thighs. Treatment of hypospadias is surgery which is commonly performed between 6 months and 2 years of age. Surgery for hypospadias repair can redirect the urethra and create a normal urinary opening on the glans. Complications are infrequent and include formation of a fistula (a hole in the repair).

Teaching video link: <https://youtu.be/K9cWLS6GQHM>



Figs. 4A and B: A type of hypospadias with the meatal opening at the underside of the penis and the hood due to absence of a part of the foreskin.

Drugs Exposure to Mother in Antenatal Period Affecting Fetal Kidney

Q8

What painkillers can be given to a pregnant woman with severe backache? Can we use angiotensin-converting enzyme (ACE) inhibitors for blood pressure control during pregnancy?

Paracetamol can be given to the pregnant lady for backache. Use of *nonsteroidal anti-inflammatory drugs such as ibuprofen, indomethacin, and nimesulide should be avoided*. ACE inhibitors as antihypertensive agents have to be *avoided*. Their use has been associated with renal tubular dysgenesis (RTD) in the fetal kidney. RTD is a potential lethal developmental anomaly of the fetal kidney. It is characterized by oligohydramnios in the mother, soft skull bones, and Potters' sequence (flattened face with large and flattened low-set ears and abnormally placed limbs) with or without pulmonary hypoplasia, and reduced or absent urine formation in the newborn. Hence, a pregnant lady should always check medication with a qualified physician before its consumption.

Single Kidney

Antenatal/postnatal scan shows single kidney with normal structure.

Q9A

Doctor, should I be worried about my child having single kidney?

Although most people are born with two kidneys, some may be born with single kidney (1 in 750 birth, more common among males and more likely left kidney to be absent). Sometimes even though one might have originally two kidneys, one of the kidneys may fail to grow properly and involute with time, and may not be seen in ultrasound scan. Functional and anatomical assessment for size and location of your child's single kidney may require some additional tests. If your child's single kidney is functioning optimally, your child should be able to undertake normal activities albeit, he will require regular follow-up. Long-term medical follow-up is required as sometimes a child with single kidney may develop renal dysfunction. Hence, regular monitoring is strongly advised which usually entails blood pressure and urinary protein measurement. Repeated nuclear scan is not recommended. The frequency of follow-up will be decided by your treating pediatrician.

Q9B

Why did my child develop this problem?

Your child has congenital (i.e., by birth) single kidney. This is usually because one of the kidneys failed to form. As explained in Q9A, there is also a possibility that originally your child had two kidneys but one of the kidneys failed to grow properly and with time it involutes. This may be genetically mediated although often an exact cause may not be established. After a detailed history and examination of your baby your pediatrician will decide, whether any genetic work-up is needed. If deemed necessary your pediatrician might ask you and your spouse to visit a pediatric nephrologist and or genetic counselor.

Q9D

As she/he has only one kidney; after birth and as my child grows up do we need to restrict any activities?

Usually, no gross restrictions in activities are advised for a child with single kidney. In fact your child should be encouraged to live a healthy life including active participation in sports and other physical activities.

Q9C

What steps do we have to take after birth to ensure that my baby stays well?

Initially, your child might have to undergo some tests. This will include a repeat ultrasound scan to properly document shape, size, and texture of the single kidney. A “nuclear” scan may also be prescribed to establish whether the nonvisualized kidney is hiding in some unusual position. Nuclear scan will also document the current functional status of the single visualized kidney. Urine examination is also advised to rule out any concurrent urinary tract infection as well as to note any presence of protein in urine. Blood tests are usually not done routinely, and are reserved for those where results of previous investigations have highlighted any concerns. If the above tests confirm normal functioning single kidney then you will be advised for regular clinic follow-up of your child. During follow-up visits, the medical staff will check blood pressure, urinalysis (check for leaking protein in the urine) as well as document appropriate growth of the single kidney through sequential ultrasound scan (There are established norms for the size of one’s kidney as per age and height. Ideally, your child’s single kidney should be larger than a normal kidney for her/his height and age as it must compensate for the absent kidney). Follow-up is usually lifelong with frequency being decided by your treating doctor. Children are encouraged to have a healthy lifestyle. Once on solid food, they are advised to have regular intake of fruits and leafy vegetables, and avoid excessive salts and junk food along with regular physical activity. You will also be advised to be alert for any urinary tract infection (UTI). Hence, if your child has fever (>100°F) without focus, then an urgent urine examination will be required, and you need to consult your pediatrician.

Multicystic Dysplastic Kidneys

Q10A

My 6-month-old son was found to have palpable right flank lump. On further evaluation, the doctor has stated that he has a nonfunctioning multicystic dysplastic right kidney (MCDK). What is MCDK and why did it occur in my child?

Multicystic dysplastic kidney (MCDK) is an abnormal development of the kidney where in the affected kidney is nonfunctioning and is replaced entirely with fluid-filled sacs or cysts (**Fig. 5**). MCDK happens in 1 in 4,000 births and is more common among boys. It is usually detected on one side as bilateral MCDK is incompatible with life. Rarely, MCDK may be a part of constellation of various problems (i.e., syndromes), which are mediated through some genetic/hereditary defects. Your pediatrician should be able to guide you regarding necessity of a genetic consultation or review by pediatric nephrologist.

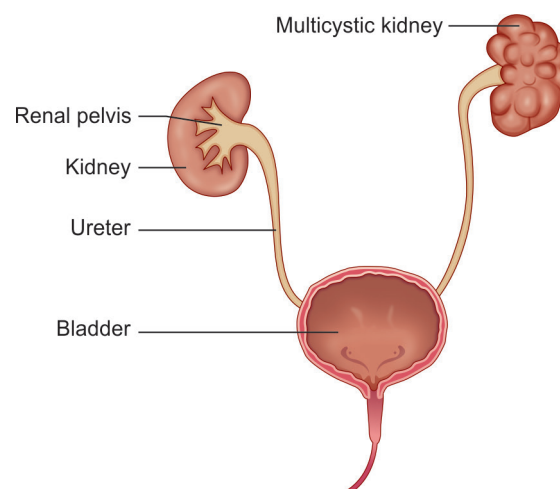


Fig. 5: Left multicystic dysplastic kidney.

Q10B**Will my child need surgery for multicystic dysplastic kidney (MCDK)?**

Multicystic dysplastic kidney usually does not cause problem as the affected kidney shrinks and disappears with time. Since the normal kidney takes over the work of both kidneys it usually becomes a bit larger than a usual kidney. Previously, MCDK would be surgically removed as it was thought to be associated with hypertension and malignancy. These risks have been found to be minimal and surgical removal is usually restricted to rare cases wherein the MCDK fails to decrease appropriately with time.

Q10C**How long do we need to keep my child under medical follow-up and what tests he will need regularly?**

Provided a good ultrasound has been undertaken nuclear imaging is usually not necessary. If there is doubt about the diagnosis, then your pediatrician might ask you to undertake a “nuclear” scan. This scan will confirm a nonfunctioning kidney which along with presence of multiple cysts in the kidney is confirmative of MCDK. This special scan will also confirm normal functioning of other kidney. MCDK is sometimes associated with problems in the other kidney (dilated collecting system with or without reflux, i.e., urine returning from the bladder to the kidney). Despite this, further test of the normal kidney is only advisable, if any abnormality is detected in that kidney on imaging (ultrasound and nuclear scans), or if your child develops infection in her/his urine and your pediatrician thinks it might be due to reflux. If the initial scans confirm a normal functioning contralateral kidney, investigations are limited to follow-up ultrasound scan to ensure that the affected kidney is shrinking, and the unaffected kidney is increasing in size and is larger than usual. Repeated nuclear scans are usually not recommended. Follow-up is lifelong and is inline for those advised for a single kidney with emphasis on regular checkup of blood pressure and urinary protein.

Horse-shoe Kidneys

Q11

Doctor, my child has been diagnosed to have “horse-shoe” kidney. What does this mean and what needs to be done?

Horse-shoe kidney happens in 1 in 500 children wherein the lower poles of the kidney might fuse together and take U-shape which mimics a horse-shoe (**Fig. 6**). Kidneys start their development in fetal stage usually in the pelvis and thereafter ascend to take up its final position in the loins. Fusion of the lower pole prevents this and the kidneys usually remain in a lower position. Horse-shoe kidney may not *per se* cause major problem. Sometimes, it is associated with dilatation of the urinary tract, vesicoureteric reflux (urine going reverse way from bladder to kidney), recurrent urinary tract infection, and rarely some part of it may be malformed carrying dysplastic (abnormal) kidney tissues. They are also associated with increased incidence of kidney stones and blood in urine (hematuria). In accordance to their position, they are situated closer to the front of the body, and are hence liable to get injured more easily than the normally placed kidneys in the loin. Initial management usually includes ultrasound scan with or without nuclear scan which usually delineates the anatomy, and function of the kidneys including their split function as well as identify any dilatation of the urinary tract. MRI of the abdomen may be rarely required to delineate the anatomy. Absence of any other abnormalities on the scans usually entails a regular follow-up to ensure normal growth of the kidneys.

N.B.: One can have kidneys not present in their normal position; these are called “ectopic” kidneys. Ectopic kidneys may have reduced function, but as long as the contralateral kidney has a normal function, it is not of much worry. Ectopic kidney may also be at higher risk of injury, if placed in less protected area but similar to single kidney or horse-shoe kidneys participation in sports activity is not contraindicated.

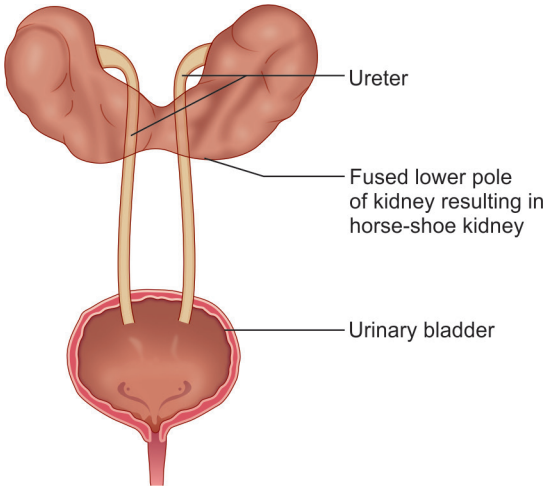


Fig. 6: Horse-shoe kidneys.

Congenital Anomalies of Kidney and Urinary Tract