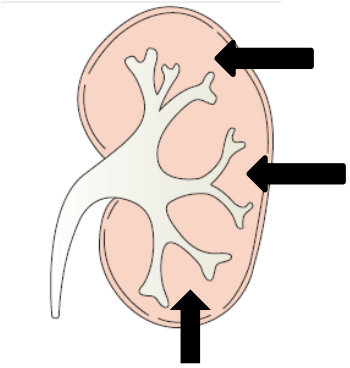
CONGENITAL ABNORMALITIES OF THE KIDNEYS

SURGICAL ANATOMY

The parenchyma of each kidney usually drains into seven calyces, three upper, two middle and two lower calyces. Each of the three segments represents an anatomical and physiologically distinct unit with its own blood supply.



The arrangement of renal calyces

Congenital abnormalities of the kidneys (Table below)

Absence of one kidney

- The prevalence of unilateral renal agenesis is about 1:1400 and it is usually discovered incidentally.
- The ureter maybe absent or;
- The ureter and renal pelvis are present but the kidney is absent.
- In either case, the solitary contralateral kidney is likely to be hypertrophied.

Table Congenital abnormalities of the kidney and ureter

Renal agenesis	
Renal ectopia	Pelvic kidney
	Horseshoe kidney
	Crossed dystopia
	Infantile polycystic disease
	Unilateral multicystic dysplastic kidney
Aberrant renal vessels	Multiple renal arteries and veins
Duplication	Duplex kidney
	Duplex renal pelvis
	Duplex kidney and ureter
Others	Congenital hydronephrosis
	Retrocaval ureter
	congenital megaureter

Absence of one kidney (box)

■ About 1:1400 people has an absent kidney

■ It is usually symptomless and often discovered by accident

■ When planning a nephrectomy the surgeon should consider the possibility that the other kidney is congenitally absent

Renal ectopia

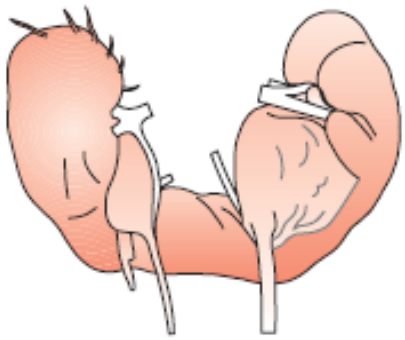
- Occurs in 1:1000.
- Ectopic kidneys are usually found near the pelvic brim and are usually left sided.
- The contralateral kidney is generally in its normal position.
- Disease in an ectopic kidney may present diagnostic problems and an unwary surgeon may be tempted to remove it as an unexplained pelvic mass.

Horseshoe kidney

- Here a pair of ectopic kidneys usually fused at their lower poles, lying in front of the fourth lumbar vertebra and great vessels.
- Horseshoe kidney is found in 1:1000 necropsies and is commoner in men.
- Upper pole fusion is rare.

Clinical features

- Horseshoe kidneys are liable to disease, possibly because the ureters angulate as they pass over the fused isthmus.
- This may lead to urinary stasis with consequent infection and nephrolithiasis.
- Sometimes there is a true pelviureteric junction obstruction, which has the same consequences.
- Horseshoe kidney is usually a radiological diagnosis.
- The most frequent appearance on the urogram shows the lower pole calyces on both sides being directed towards the midline. More rarely, all or most of the calyces are reversed.
- Horseshoe kidney is not a contraindication to pregnancy but urinary complications are more frequent.
- Division of the isthmus between the kidneys is usually only indicated in the course of surgery for abdominal aortic aneurysm.
- Although the tissue is often less vascular than normal renal parenchyma, the vascular supply of the horseshoe is typically eccentric, springing unpredictably from adjacent major vessels.

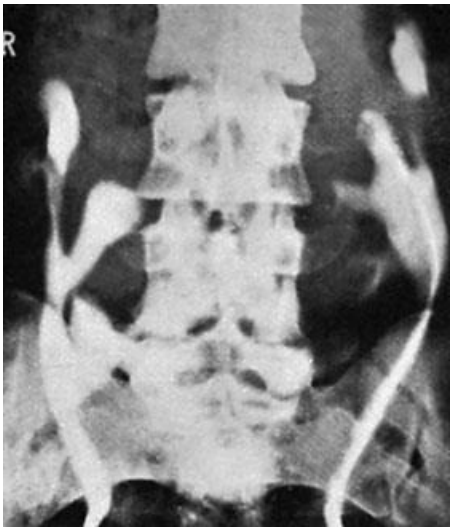


Horseshoe kidney. Note the ureters passing in front of the fused lower poles.

Horseshoe kidneys (Box)

■ Horseshoe kidneys are liable to pelviureteric obstruction, infection and stone formation

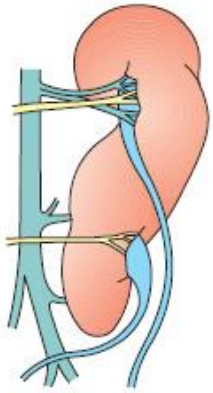
■ If not recognised, a pelvic kidney may cause diagnostic confusion during surgery



Urogram of a horseshoe kidney. Only rarely are all the calyces directed towards the spinal column

Unilateral fusion (crossed dystopia, cross fused renal ectopia)

- Unilateral fusion is rare.
- Both kidneys are in one loin and are usually fused.
- The ureter of the lower kidney crosses the midline to enter the bladder on the contralateral side.
- Both renal pelves can lie one above each other medial to the renal parenchyma (unilateral long kidney) or the pelvis of the crossed kidney faces laterally (unilateral S-shaped kidney).



Unilateral S-fusion of the kidneys

Congenital cystic kidneys (polycystic kidneys)

- Congenital cystic kidneys are hereditary and potentially lethal, and can be transmitted by either parent as an autosomal dominant trait.
- Thus, the risk of an offspring inheriting the condition can be as high as 50% depending upon the penetrance of the gene.
- The disease is not usually detectable on standard imaging until the second and third decades of life and does not usually manifest itself clinically before the age of 30 years.

Pathology of polycystic kidneys

- The kidneys are huge: the cysts distort the renal capsule.
- The renal parenchyma is riddled with cysts of varying size containing clear fluid, thick brown material or coagulated blood.
- There may be a congenital cystic disease of the liver.
- The aetiology of all renal cysts is uncertain although theories abound.

Clinical features of polycystic kidneys in the adult

The condition is slightly more common in women than men.

There are six clinical features:

1. An irregular upper quadrant abdominal mass;
2. Loin pain;
3. Haematuria;
4. Infection;
5. Hypertension;
6. Uraemia.

Renal enlargement

- Bilateral enlargement is unmistakable.
- Unilateral renal swelling, in which one kidney contains larger cysts than the other, may be confused clinically with a cystic renal tumour.

Pain

- Pain, felt as dull loin ache, is thought to be caused by the weight of the organ dragging upon its pedicle or by stretching of the renal capsule by the cysts.
- Haemorrhage into a cyst may cause more severe pain,
- The passage of a calculus from the diseased kidney also cause more severe pain

Haematuria

- Rupture of a cyst into the renal pelvis may cause recurrent haematuria.
- Profuse haematuria is uncommon.

Infection

- Pyelonephritis is common in patients with congenital cystic kidney, presumably because of urinary stasis.

Hypertension

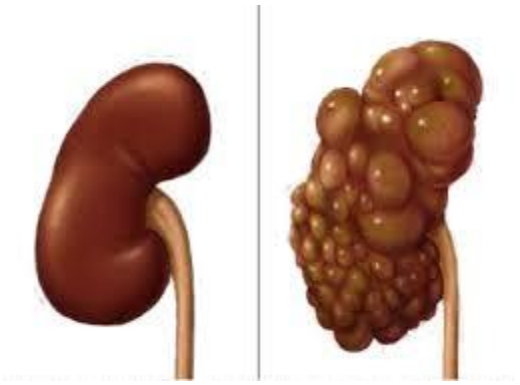
Hypertension is present in up to 75% of patients with polycystic kidneys over the age of 20 years.

Uraemia

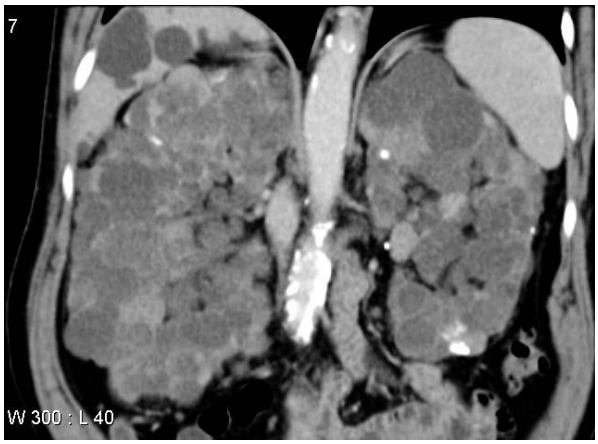
- Patients with congenital cystic kidneys pass ***large volumes of urine of low specific gravity*** containing a trace of albumin but no casts or cells.
- The non-specific symptoms of chronic renal failure develop as functioning renal tissue is replaced progressively by cysts.
- Severe anaemia is common.
- Signs of end-stage renal failure often begin suddenly during middle life and the patient is unlikely to survive without renal replacement therapy.

Imaging

- Renal imaging shows multiple cysts in both kidneys and sometimes cysts in the liver and other organs.
- Blood and debris in the cysts may mimic the heterogeneity of a cystic adenocarcinoma.
- Simple cysts are usually solitary and have smooth thin walls and homogeneous contents.
- Doubt about the diagnosis can be resolved by cytological examination of cyst fluid obtained by fine-needle aspiration.
- Polycystic kidneys have a typical appearance on urography:-
  - ✓ The renal shadows are enlarged in all directions;
  - ✓ The renal pelvis is compressed and elongated;
  - ✓ The calyces are stretched over the cysts



Left; Normal kidney
Right: Polycystic kidney



Polycystic kidney

Treatment

- ✓ As kidney failure develops, a low-protein diet will help to postpone the inevitability of renal replacement therapy.
- ✓ Infection, anaemia, hypertension and disturbances of calcium metabolism also need appropriate treatment by a nephrologist.
- ✓ Surgery to uncap the cysts (Rovsing's operation) is rarely indicated because few now accept that this can preserve renal function by relieving pressure on the parenchyma.

Polycystic kidney disease (Box)

■ Inherited as an autosomal dominant trait
■ An important cause of end-stage renal failure in adults
■ Pain, haematuria, infection and hypertension are common
■ Liable to be fatal in early middle age

Infantile polycystic disease

- Infantile polycystic disease is a rare autosomal recessive condition.
- The kidneys are large and may obstruct birth.
- Many patients are stillborn or die from renal failure early in life.
- In some children, associated congenital hepatic fibrosis is a major problem.

Unilateral multicystic disease

- Multicystic dysplastic kidney is much more common. It presents as a mass in the flank.
- Excision is the treatment of choice. Wilms' tumour, neuroblastoma and congenital hydronephrosis are all rarer causes of flank masses in childhood.

Simple cyst of the kidney

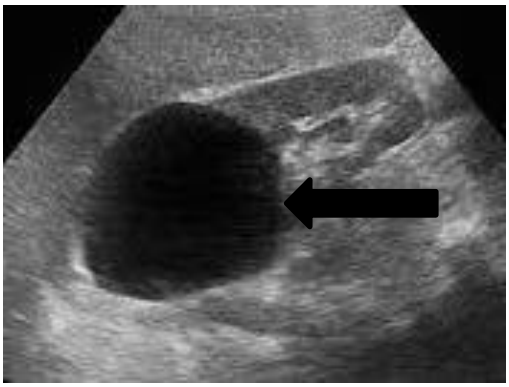
- Simple cysts in one or both kidneys are often discovered incidentally on imaging of the upper abdomen
- it is common finding
- They rarely give symptoms
- Often multiple.
- A ***palpable mass***, ***pain*** from haemorrhage into the cyst and ***infection*** are uncommon presentations.
- Rarely require treatment
- Occasionally, a cyst in the hilum of the kidney (a parapelvic cyst) presses on the pelviureteric junction and causes obstruction.
- The true nature of a cyst will be apparent by its characteristic appearance on ultrasound or computerised tomography (CT).
- Percutaneous cyst puncture for cytology is rarely necessary with modern imaging

Differential diagnosis

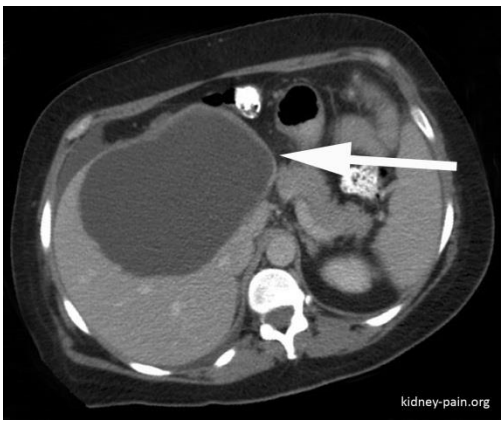
1. Hydatid cysts of the kidney.
2. On the right side; hydatid cyst of the liver.

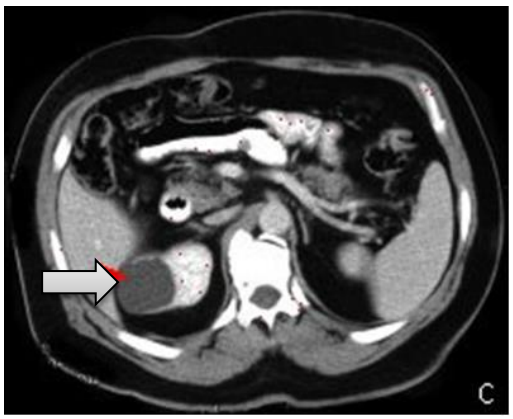
Simple renal cysts(Box)

■ Common
■ Often multiple
■ Diagnosed on ultrasound
■ Rarely require treatment
■ Treat only if causing obstruction

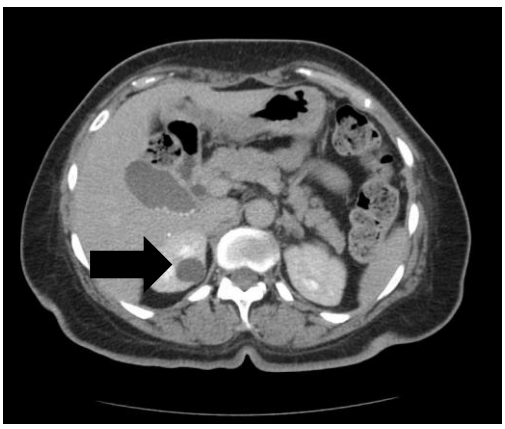


Simple cyst on ultrasound





Simple cyst on CT scan



Aberrant renal vessels

- Two or more renal arteries are most common on the left.
- The main importance of the abnormality is as a source of potential error during operations in the retroperitoneum, especially those on the kidney.
- The renal arteries are functional end-arteries, so division of an aberrant lower pole artery leads to infarction of the section of parenchyma that it supplies.
- Renal veins, by contrast, have extensive collaterals and an aberrant vein can be divided with safety.

بسم الله الرحمن الرحيم

رَبِّ اشْرَحْ لِي صَدْرِي (٢٥) وَيَسِّرْ لِي أَمْرِي (٢٦) وَاحْلُلْ عُقْدَةً مِنْ لِسَانِي (٢٧) يَفْقَهُوا قَوْلِي

صدق الله العظيم

سورة طه

د مقداد فؤاد

السلام عليك يأبا عبد الله