Wrong place, wrong time: Imaging and embryological features of congenital renal anomalies

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Learning objectives

- To understand the normal embryological development of the kidneys.
- To understand the development of congenital renal ectopia and fusion resulting from failures in embryological development.
- To be able to recognize and correctly diagnose renal fusion anomalies and ectopia on various imaging modalities.
- To appreciate some of the pathological consequences of congenital renal and renal tract anomalies.
**Background**

Congenital renal parenchymal anomalies occur in approximately 1% of the population [1]. The importance of these anomalies is two-fold. First, these anomalies should not be mistaken for pathology themselves. Second, it is important to recognise that pathology, such as infection or calculus formation, can occur more frequently in these anomalies.

Embryological development of the kidney and collecting system represents the complex interplay between two embryological structures: the metanephros and ureteric bud. Congenital renal anomalies are the result of abnormal development, abnormal migration and/or abnormal fusion.

The collecting system arises from the ureteric bud, which arises from the mesonephric duct in the fourth week of gestation. The renal parenchyma arises from the metanephros, a derivative of the intermediate mesoderm, which appears in the fifth week.

The ureteric bud penetrates the metanephric mesoderm, which forms as a cup-shaped tissue cap. The ureteric bud dilates and subdivides to form twelve or so generations of tubules, with the first generations fusing to form the renal pelvis, major and minor calyces, and renal pyramids, and the later generations forming approximately a million renal tubules. Early division of the ureteric bud can result in a duplex kidney or other anomalies of the ureter.

Under complex signalling pathways the ureteric bud incites the metanephric tissue to form small renal vesicles that eventually form primitive S-nephrons that are invaginated by endothelial cells from nearby angioblasts, and go on to form the definitive nephron. Errors in the signalling pathway or formation of the metanephric tissue cap can result in renal agenesis or other renal parenchymal abnormalities.

The kidney develops in the pelvis, but rises to its normal abdominal location in adults due to disproportionate growth of the body in the lumbar and sacral regions. During the relative ascent of the kidney, as they pass through a fork formed by the umbilical arteries, errors may result in an abnormal position or fusion.
Images for this section:

Fig. 1: Chronological diagram demonstrating the embryological development of the kidneys as they form and migrate to their final position. Note the constantly changing position of the vascular supply.

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Horseshoe kidney is the most common of all renal fusion anomalies, accounting for 90% of all such anomalies, and occur in approximately 0.25% of the population, with a predilection for males in the order of approximately 2:1 [2].

Embryology

During ascent towards the renal fossa, the foetal kidneys cross the umbilical arteries. Any aberration in position of the umbilical arteries may result in fusion of the bilateral nephrogenic blastemas. When this fusion occurs at the lower poles of the kidneys, a U-shaped single kidney is formed - resembling the shape of a horseshoe.

Imaging Findings

Horseshoe kidneys may be found anywhere along the path of normal renal migration that occurs during foetal development. They usually lie anterior to the great vessels (the aorta and inferior vena cava). However, cases have been described where they are positioned posterior to, or in between the great vessels [2]. The isthmus of a horseshoe kidney consists of either functional renal parenchyma or fibrous tissue, and is most often located inferior to the origin of the inferior mesenteric artery [2]. Fig. 2 on page 11

Blood supply is variable, with up to 30% of cases receiving blood from a single renal artery to each kidney [2]. For the remainder of cases, there may be duplicate or triplicate renal arteries to one or both kidneys, whilst the isthmus may receive blood from branches of the renal arteries or from branches arising from other nearby major vessels, such as the aorta, inferior mesenteric artery and iliac arteries [2].

The inferior pole of each kidney points medially, which results in the ureters passing anterior to the isthmus. This abnormality of renal axis orientation leaves the pelvicalyceal system of horseshoe kidneys prone to obstruction, which results in a higher risk of infection, hydronephrosis and calculus formation.

Key Points

• A single horseshoe-shaped kidney
• Results from fusion of the lower poles
• Located anywhere along the path of normal renal migration, but usually lower than normal, ascent impeded by the inferior mesenteric artery
Variable blood supply

**Pancake kidney**

Pancake kidney is a rare fusion anomaly that occurs in approximately 0.04% of the population [3]. Also known as 'cake kidney', 'disc kidney', 'lump kidney' or 'fused pelvic kidney', the defining characteristics are that of a single fused renal mass that lies within the pelvis. Fig. 3 on page 11

**Embryology**

The anomaly results from fusion of both nephrogenic blastemas with early arrested renal migration during foetal development. Migration is arrested whilst the fused renal mass is still within the pelvis during embryogenesis, preventing the normal cephalad ascent of the kidneys. During this ascent, the blood supply is derived from nearby major arteries and is re-established progressively as the renal masses reach their final position. Hence, in pancake kidney, the blood supply reflects the fact that renal migration has been stopped. The single renal mass receives blood from branches of nearby major arteries, such as the lower abdominal aorta or common iliac arteries, which increases the risk of haemorrhage in cases of pelvic trauma.

**Imaging Findings**

The shape of a pancake kidney is non-reniform, lobulated and often flattened. Commonly two short ureters are present, which arise from an anteriorly positioned collecting system and enter the bladder in a normal relationship; however, cases have been described where only a single ureter is present [3]. A pancake kidney may be located in the presacral or prevertebral space, or at the level of the aortic bifurcation.

Concomitant anomalies in other extrarenal organs such as: tetralogy of Fallot, vaginal absence, sacral agenesis and spina bifida have been reported in many diagnosed cases of pancake kidney [3]. This highlights the importance of screening for such associated abnormalities when the diagnosis is made.

**Key Points**

- A single fused renal mass in the pelvis
- Results from abnormal fusion of the kidneys and failure to ascend
- Blood supply from foetal vessels originating from local major arteries
- Concomitant congenital abnormalities in other organs

**Cross fused ectopia**
Cross fused ectopia is the second most common fusion anomaly after horseshoe kidney, with an estimated incidence of up to 0.08% [4]. It is both a fusion and ectopic anomaly characterised by one kidney that has crossed the midline and fused with the kidney on the opposite side.

**Embryology**

The aetiology of cross fused ectopia is not exactly known. Proposed mechanisms include: effect of teratogens, a wandering ureteral bud causing nephrogenesis in an atypical location, abnormally located umbilical arteries hindering the normal cephalad migration of the kidneys causing one to cross the midline along the path of least resistance, and aberrant development of the caudal end of the foetus with malrotation [4].

**Imaging Findings**

The most common configuration is that the left kidney is ectopic and located on the right, with fusion of it's upper pole to the lower pole of the normally positioned right kidney [4]. As a result, the ureter arising from the ectopic kidney crosses the midline and enters the bladder in a normal relationship. Fig. 4 on page 12

The arterial supply and venous drainage of this anomaly is usually grossly abnormal with a variety of possibilities. The ectopic kidney usually receives blood from a vessel on the ipsilateral side, however cases have been described where the arterial supply originates on the contralateral side. There can be up to a total of six major arteries supplying the fused renal mass, with 25% of them receiving blood from branches of the upper abdominal aorta and the remainder of cases receiving blood from branches of either the lower abdominal aorta or the iliac arteries [4].

As with other fusion and ectopic abnormalities, there tends to be a higher risk of developing hydronephrosis, renal calculi and infection. Such complications may be the reason a patient is imaged and the diagnosis made, however a significant proportion of cases are asymptomatic and therefore diagnosed incidentally during imaging for other indications.

**Key Points**

- A single fused renal mass located on one side of the body
- Two ureters, with one crossing the midline
- Blood supply grossly abnormal

**Renal Agenesis**
Renal agenesis is the absence of renal tissue due to failure in embryogenesis, and can occur unilaterally or bilaterally. Bilateral renal agenesis is a fatal condition that may result in miscarriage or death in the early neonatal period. It is one of the causes of oligohydramnios, which results in Potter sequence.

Unilateral renal agenesis is also known as solitary kidney and has an incidence of approximately 1 in 1000 to 1 in 2000 newborns [5].

**Embryology**

The aetiology in many cases of renal agenesis is currently unknown, but is thought to be multi-factorial. An early vascular insult to the developing ureteric bud has been proposed.

Embryologically, renal agenesis results from a failure of the proper development of the metanephros resulting in complete absence of a renal structure.

**Imaging Findings**

In unilateral renal agenesis, the contralateral kidney is often hypertrophic as a compensatory mechanism. There is usually no renal vein, artery or ureter on the affected side, although in some cases a fibrotic cord may be seen in lieu of the ureter. This cord will usually end ectopically rather than enter the bladder [5]. The adrenal gland of the affected side is present in 90% of cases [5]. Fig. 5 on page 13

Unilateral renal agenesis is associated with other congenital anomalies of the kidneys and urinary tract in approximately 32% of cases, most commonly vesicoureteral reflux[6]. Extra-renal anomalies are found in up to 31% of cases [6]. Hence, the early diagnosis of a solitary kidney is important as the condition requires clinical follow-up.

**Key Points**

- Absence of one kidney
- Fibrotic cord corresponding to the ureter may be seen
- Absence of the corresponding renal artery and vein
- Often associated with other renal, extra renal and urinary tract abnormalities

**Pelvic kidney**

This is an ectopic anomaly, where one kidney remains fixed retroperitoneally within the bony pelvis.

**Embryology**
The ectopia arises from a failure of the kidney to ascend cranially during embryogenesis, hence ectopic kidneys may be found anywhere along the pathway of normal renal ascent - such as the pelvis and lower abdomen. In rare cases, ectopic kidneys may be found in the thorax due to excessive cranial migration prior to diaphragmatic closure.

**Imaging Findings**

Ectopic kidneys are often small, lobulated, malrotated and can have extrarenal calyces. Their blood supply is usually derived from surrounding vessels, such as the iliac arteries. Typically, patients with pelvic kidney(s) are asymptomatic and the diagnosis is usually made incidentally during imaging for other indications. Due to the ectopic location of the kidney, referred pain caused by pyelonephritis or calculi may be atypical, and patients may be misdiagnosed, for example with acute appendicitis. Fig. 6 on page 14

Complications such as vesio-ureteric reflux or pelvi-ureteric junction (PUJ) obstruction are common. These complications may be a consequence of a malrotated kidney or abnormal blood supply which obstructs the collecting system. The ectopic location and blood supply present a treatment challenge in regards to surgery or trauma in the pelvis as the risk of injury to aberrant vessels, overlying nerves and viscera is increased.

**Key Points**

- One kidney remains in the pelvis, whilst the other is normally located
- Results from a failure to ascend during embryogenesis
- Abnormal blood supply from surrounding vessels
- High incidence of vesico-ureteric reflux and PUJ obstruction

**Duplex kidney**

Duplication of the renal collecting system is the most common congenital abnormality of the urinary tract, with an estimated incidence of up to 5% [7]. The abnormality may be complete, in which the affected kidney has two pelvicalyceal systems, with two ureters that enter the bladder separately. Partial duplication results in two pelvicalyceal systems with either a single ureter draining both, or a bifid ureter that joins distally to enter the bladder via a single orifice. Of the two, partial duplication is more common and accounts for over 95% of cases [7].

**Embryology**

Duplication anomalies occur very early during embryogenesis. Complete duplication is thought to arise as a result of two ureteric buds which join with the metanephric blastema, whilst partial duplication is thought to result from a single ureteric bud that bifurcates prior to it joining the metanephric blastema.
Imaging Findings

In a complete duplication, the upper pole renal moiety is drained by its own single ureter, whilst the lower pole moiety has its own single ureter. The position of the insertion sites of the distal ends of the ureters into the bladder have been observed to be nearly constant and are described by the Weigert-Meyer rule, which states that the ureter draining the lower pole moiety enters the bladder in an orthoptic location that lies more lateral and cephalad, whilst the ureter draining the upper pole moiety enters the bladder in an ectopic location that is more medial and caudal and most often ends in a ureterocoele. There have only been a few rare reports of cases where there are exceptions to this rule [7].

The consequence of the Weigert-Meyer rule is that the ureter draining the lower pole moiety is prone to vesico-ureteric reflux, whilst the ureter draining the upper pole moiety is prone to obstruction. Fig. 7 on page 15

Key Points

- Two pelvicalyceal systems draining a single kidney
- Single bifid ureter, or two separate ureters
- Normal renal vascular supply and drainage
- Complete duplication follows the Weigert-Meyer rule
Fig. 2: A. Illustration of the typical appearances of a horseshoe kidney, with the isthmus below the inferior mesenteric artery and anterior facing renal pelvises. B. Ultrasound scan of a horseshoe kidney demonstrating the isthmus. C. Axial T2 weighted magnetic resonance image of the abdomen demonstrating a horseshoe kidney with the isthmus passing anterior to the aorta and inferior vena cava. D. Intravenous pyelogram (IVP) demonstrating a horseshoe kidney with contrast in both inferior poles tracking towards the midline, creating a horseshoe configuration. E. Volume rendered three dimensional computed tomography urogram demonstrating typical appearance of a horseshoe kidney. F. Dimercaptosuccinic acid scan(DMSA) of the kidneys demonstrating marked tracer uptake medial to both inferior renal poles within a functional isthmus confirming the diagnosis of horseshoe kidney.

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**Fig. 3:** Illustration of a pancake kidney with a pelvic location, with the vascular supply originating from the common iliac arteries and anterior facing renal pelvises.

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Fig. 4: A. Illustration of some of the various configurations that may be observed with a cross-fused anomaly. The illustrations depict (left to right): anterior facing renal pelvises with the upper moiety draining to the contralateral side without crossing ureters, anterior facing pelvises with the lower moiety draining to the contralateral side with crossing ureters, a sigmoid kidney, and a side-by-side fusion. B. Coronal reformatted contrast enhanced CT abdomen (portal venous phase) demonstrating a left cross fused renal ectopic anomaly, with absence of a kidney in the right renal bed. The renal conglomerate on the left consists of a normal sized kidney with a smaller, mal-positioned kidney fused superior to it with it's own renal pelvis. This patient also has duplication of their inferior vena cava. C. An IVP demonstrating right sided cross fused renal ectopia. Contrast can be seen filling two renal pelvises on the right, with two separate ureters draining the fused renal mass, and absence of the left renal outline. D. 3D volume rendered image from a contrast enhanced CT scan (renal excretory phase) showing a unilateral renal mass with two collecting systems. The caudal collecting system drains to a ureter that crosses the midline to insert in the normal anatomic location on the contralateral side. E. Diemercaptosuccinic acid (DMSA) scan of a child, demonstrating unilateral tracer uptake in a large left kidney with two distinct segments that appear to be fused.

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Fig. 5: A. Absent kidney in the right renal bed (arrow) B. Contrast enhanced CT scan (renal parenchymal phase) showing an absent kidney on the left, with a large right kidney due to compensatory hypertrophy. C. 3D volume-rendered image of the same same patient as in (B) showing an absent left kidney.

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Fig. 6: A. PET scan of a patient with a left colonic primary tumor and extensive peritoneal metastases. A left pelvic kidney was incidentally noted. B. Coronal reformatted image of a contrast-enhanced abdominal CT scan (renal excretory phase) showing the left kidney immediately superior to the bladder near the midline. C. 3D volume-rendered image of the same patient as in (B).

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Fig. 7: A. Illustration depicting the Weigert-Meyer rule, with the upper moiety entering the bladder in an ectopic location inferior and medial to the normal anatomic insertion of the lower pole moiety. B. Fluoroscopic micturating cystourethrogram (MCUG) demonstrating a duplex collecting system on the left, with grade 5 reflux into both moieties. The open arrow indicates the upper moiety and the closed arrow the lower. C. An IVP showing bilateral duplex collecting systems, with a ureterocele on the left (arrow) demonstrating the 'cobrahead sign', illustrating the typical appearances of the Weigert-Meyer rule. D. Ultrasound image of the right kidney showing two echogenic complexes with intervening cortical tissue creating a typical appearance of a duplex kidney. There is no evidence of obstructive hydronephrosis in either moiety. E. An IVP of a patient with a right sided duplicated collecting system. In this case the upper moiety is obstructed and non-functioning, displacing the lower pole moiety inferiorly. This creates the appearance of a drooping lily in the lower moiety, and thus the 'drooping lily sign'. F. 3D volume-rendered image from a contrast-enhanced CT scan (renal excretory phase) showing bilateral completely duplex collecting systems with normal drainage. The left kidney has two incidental simple renal cysts.

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Conclusion

The array of congenital renal anomalies is vast and complex. Fusion and ectopic abnormalities are the result of aberrant fusion or migration of the kidneys during foetal development. Correct diagnosis relies on understanding the normal embryological development of the kidneys and recognition of anomalies when this normal development has been altered.

Most congenital renal and urinary tract abnormalities are asymptomatic and incidentally discovered, however they often put the patient at higher risk of developing significant pathological complications whose consequences can be minimised by early diagnosis and treatment. Thus, the recognition and correct radiological diagnosis of a congenital renal or urinary tract abnormality is paramount to the long term care of such patients.
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