Ultrasonography (US) is a modality of choice to evaluate the fetal renal anomalies, but making a precise prenatal diagnosis may be difficult or impossible, especially during the early second trimester due to fetal position, maternal obesity, superimposed bone, and in particular, oligohydramnios. Magnetic resonance imaging (MRI) is gaining more and more popularity as a complementary modality because it images the fetus in multiple planes, a large field of view, using excellent tissue contrast, and irrespective of fetal position [1-6]. Congenital renal anomalies include renal dysgenesis (agenesis and hypoplasia), congenital cystic renal disease, cystic renal disease associated with other syndromes, hydronephrosis, congenital renal positional anomalies, and congenital renal morphological anomalies [2, 5-7]. In this review, fetal renal MRI technique is discussed, and the congenital renal anomalies observed on MRI are illustrated.

MRI Technique

MRI was performed using a 1.5 T MR scanner (Twin-speed, GE Medical Systems, Milwaukee, WI) with a torso phased-array coil, and images were sequenced at a single-shot fast-spin echo (SSFSE) and thick-slab T2-weighted sequences. The imaging protocol comprised SSFSE sequences in the axial, coronal, and sagittal planes of the fetal kidneys (TE: 90 ms, TR: minimum, field of view: 270 × 270 mm, matrix: 256 × 224, slice thickness: 4 or 5 mm, scan time: 34 s/19 slices) and thick-slab T2-weighted sequences in the coronal and sagittal planes (TE: minimum, TR: 4000 ms, field of view: 300 × 300 mm, matrix: 448 × 224, slice thickness: 60 mm, scan time: 33 s/9 slices). If necessary, one could serially repeat scans of multiple sections (axial, coronal or sagittal), which might be expected to improve the rate of detection of the tiny or complicated lesions. All sequences were performed without or with maternal breath-holding. The entire examination remained within 30 min.

Normal Fetal Kidneys on T2-Weighted Images

The renal parenchyma is brighter than fetal liver, spleen, or muscle, but less bright than maternal fat on SSFSE T2-weighted images. The renal pelvis appears...
bright but is isointense to maternal fat, and represents fetal urine. Occasionally, a long-T2 rim surrounds the kidney, suggesting perirenal fat (Fig. 1). On thick-slab T2-weighted images, a topographic view of the normal bulk fluids within the spinal canal, lungs, stomach, intestines, and urinary tract, including bilateral renal pelves and urinary bladder, is visible (Fig. 2).

**TYPES OF FETAL RENAL ANOMALIES**

**Renal agenesis**

Renal agenesis, congenital absence of one or both kidneys, possibly results from a failure of the ureteral bud to induce development of the metanephrogenic blastema. Bilateral renal agenesis is a rare and devastating anomaly, occurring in only one or two per 10,000 births; the condition is usually associated with severe oligohydramnios and is one causative agent of Potter sequence [8]. Unilateral renal agenesis is much more common and usually found incidentally along with compensatory hypertrophy of the contralateral kidney, or may be associated with ipsilateral urogenital anomalies or adrenal agenesis [8, 9]. On MRI, anhydramnios or oligohydramnios, bowel loops in place of normal kidney in the expected renal fossa, and a small or signal void of the urinary bladder [5] suggest bilateral renal agenesis (Fig. 3). In a fetus with unilateral agenesis, absence of the bright urine signal from the involved renal pelvis, contralateral renal hypertrophy, normal visualization of the urinary bladder and a normal amniotic fluid volume are observed (Fig. 4) [3, 5, 10, 11].

**Renal hypoplasia**

Renal hypoplasia refers to a small kidney containing essentially normal residual parenchyma with intact, but fewer nephrons, whereas a dysplastic kidney contains disorganized and maldifferentiated tissue [12]. In practice, the diagnosis of congenital renal hypoplasia is favored under the following conditions: the renal size is decreased by two standard deviations from the mean size according to age and the contralateral kidney shows compensatory hypertrophy; renal scarring is excluded using postnatal Doppler US as an initial renal screen or 99mTc–dimercaptosuccinic acid radionuclide scan [12]. Renal hypoplasia is not generally associated with urinary tract malformations but may be a component of a larger genetic syndrome such as renal coloboma and branchio-oto-renal syndromes, though it may be an isolated finding [13]. On MRI, the abnormal small kidney maintains a normal renal pelvis (Fig. 5).

**Congenital cystic renal diseases**

Congenital cystic renal disease can develop in the fetus but more typically develops after birth. The condition has two MRI patterns, frank cysts and long-T2 renal parenchyma, which can be associated with each other. The cysts can be miniscule or large, and can develop anywhere in one or both kidneys. Long T2 of the renal parenchyma suggests microscopic cysts and/or dilated tubules. The Potter

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**Figure 1**

Axial fetal MRI at 22 weeks of gestation. The renal parenchyma (arrows) appears less bright than the maternal fat (F). The renal pelvis is bright and isointense to the maternal fat, indicating fetal urine. The bright signal surrounding the kidney suggests perirenal fat (arrowheads).

**Figure 2**

Thick-slab fetal MRI at 27 weeks of gestation. A global view reveals the normal static fluids in the spinal canal, lung (L), stomach (S), bowel loops (arrows), renal pelvis (arrowhead), and urinary bladder (B).
classification of cystic renal disease [14, 15] comprises four types: type 1, infantile polycystic kidney disease (autosomal recessive polycystic kidney disease [ARPKD]); type 2, cystic dysplastic kidney disease (multicystic dysplastic kidney disease [MCDK]); type 3, adult polycystic kidney disease (autosomal dominant polycystic kidney disease [ADPKD]); and type 4, obstructive cystic renal dysplasia.

**Type 1: Autosomal recessive polycystic kidney disease**

ARPKD is the most common heritable cystic renal disease in infant and children, occurring at a frequency between one in 6,000 to 55,000 births [16, 17]. The disorder

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**Figure 3.** Bilateral renal agenesis in a 34-week-old fetus. **a.** Axial and **b.** coronal MRI shows oligohydramnios, a signal void at the urinary bladder (arrowhead), and neither kidney is visible in the expected renal fossa, instead replaced by small bowel (arrows) and colon loops (C).

**Figure 4.** Right renal agenesis in a 25-week-old fetus. Coronal MRI shows an absent urine signal in the right renal pelvis, normal left renal contour, visible urinary bladder (not shown), normal lung development, and normal amniotic fluid volume.

**Figure 5.** Right renal hypoplasia in a 23-week-old fetus. Axial MRI shows a small right kidney (arrows) compared to left kidney and a normal bright urine signal in the bilateral renal pelves.
manifests as nonobstructive dilatation of collecting tubule or renal medulla ectasia, and is typically bilaterally symmetric. In severe cases, these 1-2 mm diameter lesions may extend to the cortex. The kidneys are typically enlarged when numerous ducts are involved. Patients with ARPKD can also have hepatic anomalies, including numerous, dilated bile ducts, as well as enlarged and fibrotic portal tracts. On MRI, the enlarged well-delineated kidneys with high T2 signal intensity are indicative of numerous dilated collecting ducts in the renal parenchyma; the normally bright renal pelves may be obscured as well [3, 18, 19]. Concurrent pulmonary hypoplasia and oligohydramnios can be also seen (Fig. 6) [19]. The hyperintense medullary lesions help differentiate ARPKD from other causes of renal hyperechogenicity found on US, such as ADPKD, congenital nephrosis, Bardet–Biedl syndrome, or cytomegalovirus infection [3].

Type 2: Multicystic dysplastic kidney disease
MCDK is characterized by a disturbed nephrogenic tissue differentiation. The renal parenchyma is randomly replaced by variably sized non-communicating cysts resembling bunches of grapes; ultimately, the reniform shape of the kidney is completely lost [14]. Segmental MCDK may occur in one pole of a duplex kidney or within a crossed fused ectopic kidney. The abnormality may be unilateral (76%) or bilateral (24%) and occurs more frequently in males, at 2.4:1 [7, 20]. In unilateral cases, oligohydramnios is absent, and the bladder appears normal [21]. However, in bilateral cases, there is severe oligohydramnios, and the urinary bladder is not visible; Potter’s sequence usually results [20]. The MRI shows an enlarged lobulated kidney with randomly scattered variably sized cysts [5] resembling bunches of grapes (Fig. 7). In severe cases, the kidneys appear as large abdominal cystic structures [22, 23].

Type 3: Autosomal dominant polycystic kidney disease
ADPKD is a dominant heterogenetic systemic disease,
affecting approximately one in 500-1000 births [24, 25]. It is characterized by focal and progressive enlargement of renal cysts, and as a systemic disorder, multiple organs are affected, resulting in extrarenal cysts in the liver, pancreas, spleen, thyroid, arachnoid membranes, and macrovessels. ADPKD is genetically heterogeneous and is attributed to mutations in PKD1 (chromosome 16), which is more associated with renal disease, and PKD2 (chromosome 4), which causes extrarenal manifestations such as intracranial aneurysms [15, 25, 26]. In utero, only the renal lesions can be detected by US [15] or MRI. Increased renal echogenicity is the most common US finding in ADPKD, and identification of simple cysts is rare in utero (1:1100 pregnancies). Although cysts are the diagnostic criteria for ADPKD, cystic changes seldom occur in fetus and are found predominantly in the cortex [15, 26]. In MRI, both ARPKD and ADPKD demonstrate high T2 signal intensity in utero; however, the kidney tends to be larger and have a brighter T2 signal in individuals with ARPKD compared to those with ADPKD. Besides ARPKD, other differential diagnoses include Bardet–Biedl syndrome, Meckel–Gruver syndrome, and Ivemark II syndrome [26]. Diagnosis of ADPKD is supported by the following findings: high T2 renal signal intensity with a normal renal pelvis and tiny subcapsular cysts [15]; the absence of concurrent malformations such as polydactyly, spinal abnormalities, hepatic fibrosis, and central nervous system anomalies; normal amniotic fluid volume; visible urinary bladder; and normal lung development (Fig. 8).

Type 4: Obstructive cystic renal dysplasia

This condition is defined as an abnormal nephronic and ductal development due to urinary tract obstruction during the early first trimester such as ureteropelvic junction obstruction, vesicoureteral junction obstruction, vesicoureteral reflux, urethral agenesis, and posterior urethral valves [14, 27]. In utero, obstruction during the first trimester causes cystic renal dysplasia, whereas obstruction during the second trimester causes hydronephrosis or atrophy [27, 28]. The MRI shows a normal to small renal size, preserved reniform shape, and scattered small cysts associated with urinary tract obstruction (Fig. 9). Differential diagnoses include MCDK (usually enlarged lobulated kidney with larger cysts and no urinary tract obstruction) and Meckel–Gruver syndrome (concurrent occipital encephalocoele and polydactyly).

Cystic renal diseases within syndromes or sequences (glomerulocystic and medullary cystic dysplasia)

Many syndromes or sequences are also associated with renal cysts. Distinguishing between glomerulocystic and medullary forms requires pathologic examination. In glomerulocystic kidney disease, the cysts result from dilation of the Bowman’s space; subcapsular cysts are diagnostic on MRI or US. The disease is rare and may occur in otherwise normal infants or associated multiple malformations such as ADPKD glomerulocystic type, oral facial digital syndrome, short rib-polydactyly syndromes, Trisomy 13, Trisomy 18, tuberous sclerosis complex, Jeune syndrome (asphyxiating thoracic dysplasia), Zellweger syndrome.

Figure 8. ADPKD in a 33-week-old fetus. Coronal MRI shows a bilateral high renal signal intensity, normal renal pelvis brightness, tiny subcapsular cysts (arrows), normal amniotic fluid volume, and normal lung development.

Figure 9. Obstructive cystic renal dysplasia in a 28-week-old fetus caused by vesicoureteral reflux. Coronal MRI shows a normal left renal contour (arrows) with hydronephrosis and several small cysts (arrowheads).
(cerebrohepatorenal syndrome), Ivemark II syndrome, and others [29]. Medullary cystic dysplasia is characterized by extensive medullary tubule dilation extending towards the cortex but primarily affecting the medulla. Associated diseases include Meckel–Gruber, Bardet–Biedl, and Beckwith–Wiedemann syndromes [30, 31]. Similar to ARPKD, syndromes or sequences associated with glomerulocystic kidney disease and medullary cystic dysplasia may show nephromegaly with high T2 signal intensity [32] or small renal cysts [33].

**Hydronephrosis**

Fetal hydronephrosis is a common finding and occurs in 0.5 to 1 percent of pregnancies [34]. Although typically a common and transient physiologic state in most cases, it can occasionally be caused by urinary tract obstructive uropathy (ureteropelvic junction obstruction, posterior urethral valve) and non-obstructive causes (vesicoureteral reflux, megaureter, prune-belly syndrome) [35]. The most common causes are transient hydronephrosis, ureteropelvic junction obstruction, and vesicoureteral reflux. However, ureteropelvic junction obstruction and vesicoureteral reflux may manifest initially as mild hydronephrosis. In severe cases, the normal renal morphology is lost, resulting in a large, lobulated cystic structure visibly that may be difficult to distinguish from other abdominal cystic masses. On MRI, the features of ureteropelvic junction obstruction include varying degrees of pelvicaliectasis without ureteral dilatation, ectopic ureterocele, megaureter, or posterior urethral dilatation (Fig. 10). MRI also provides superior anatomic details and a large field of view to detect complex urinary tract abnormalities (Fig. 11) [36].

**Congenital renal positional anomalies**

An ectopic kidney may be pelvic, iliac, abdominal, thoracic, or crossed and is often associated with other abnormalities such as contralateral renal agenesis, vascular malformation, and genital anomalies [37]. Pelvic kidney is the most common type, while ectopic intrathoracic kidney is the least frequent, occurring in less than 1 in 10,000 births [38]. Most ectopic kidneys are clinically asymptomatic except for the vesicoureteral reflux development, hydronephrosis, urolithiasis, and increased susceptibility to injury from blunt trauma. Ectopic kidney is frequently first diagnosed on US, usually incidentally. Intrathoracic renal ectopia may be related to diaphragmatic evagination or hernia and is usually an associated finding (Fig. 12).

**Congenital renal morphological anomalies**

Horseshoe kidney is the most common renal fusion anomaly. Most cases of horseshoe kidneys are asymptomatic and found during autopsy. The condition may increase the risk of hydronephrosis, renal stones, infection, transitional cell carcinoma and other cancers, Wilms’ tumor, and carcinoid tumor; it may also accompany genitourinary and other congenital anomalies. A renal pelvic angle less than 140° on an axial US highly suggests horseshoe kidney [39].
CONCLUSIONS

MRI is a useful complementary diagnostic tool to assess fetal renal anomalies diagnosed by US. It is essential for both prenatal counseling and peri- and postnatal management to selected patients when US findings are inconclusive or inadequate, especially if oligohydramnios is present.3.

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REFERENCES

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Figure 12. Congenital diaphragmatic hernia in a 29-week-old fetus. On axial MRI, the liver (L), small bowel loops (arrows), and right kidney (arrowheads) are herniated into the right thorax, causing left displacement of the heart.


