Fetal Genito Urinary System Anomalies and Miscellaneous Conditions

15.1 Genito Urinary System Anomalies

Normal kidneys are visualized from 15weeks and appear as elliptical paravertebral organs. The parenchyma shows intermediate signal intensity and the pelvicalyceal system appears hyperintense on T2-weighted HASTE images (Fig. 15.1). The renal cortex appears mildly hypointense as compared to the medulla with the renal corticomedullary differentiation becoming more and more prominent as the gestational age advances [1, 2]. The true-FISP sequence is helpful in the better delineation of obstructive changes in the urinary tract. T1W images are useful in differentiating dilated ureters from adjacent bowel loops as meconiumfilled large bowel appear hyperintense as compared to the

X

hypointense dilated ureter [2]. Kidneys appear hyperintense on DWI due to the high cellularity (Fig. 15.2a, b). DWI are very useful in the detection of renal parenchyma and in the assessment of renal functionality. A diseased fetal kidney shows reduced brightness on DWI due to decreased cellularity. Hence, increased ADC values of renal parenchyma are indicative of impaired renal functions. MRI is also helpful in the assessment of associated VATER (vertebral, anal atresia, trachea-esophageal fistula, renal) and pulmonary anomalies. Urinary tract abnormalities can impact fetal lung maturity and thus affect the prognostic implications. MRI is complementary to US when the evaluation is hampered due to oligohydramnios, maternal obesity, and overlying bones.



Fig. 15.1 Axial (a) and coronal (b) T2W images of a 30-week fetus showing normal kidneys. K kidneys

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Fig. 15.2 Axial DW images (**a**, **b**) and axial T2W image (**c**) of a 29-week fetus showing normal kidneys. The kidneys appear hyperintense on DWI (arrows). *K* kidneys

15.1.1 Unilateral Renal Agenesis

Definition Absent kidney on one side.

Ultrasound and doppler are diagnostic. Presence of renal artery is useful to pinpoint the presence of kidney and *MRI* is rarely done.

Imaging Features

- Kidneys appear hyperintense on DWI and they are easily located on MRI.
- The ipsilateral renal fossa is empty with absent DWI hyperintense signal elsewhere in the abdomen favors agenesis.
- The contralateral kidney shows compensatory hypertrophy.
- MRI is also useful to differentiate bowel from kidney when there is doubt on sonography.
- MRI is useful when there is associated oligohydramnios [3, 4].

Differential Diagnosis Globular adrenal may mimic a kidney.

Colonic loop may mimic a kidney.

15.1.2 Ectopic Kidney

- MRI is useful to identify ectopic kidney (Fig. 15.3). It is helpful to differentiate bowel from kidney when there is doubt on sonography [5].
- The ipsilateral renal fossa is empty with the presence of extrarenal/pelvic location of DWI bright signal favors ectopia.

15.1.3 Fusion Abnormality of Kidneys

• Sometimes fusion abnormality may appear as a mass on sonography, MRI is useful to identify the abnormality.



Fig. 15.3 Coronal T2W images of a 34-week fetus showing ectopic left kidney (arrows), *K* kidneys

• Crossed fused ectopia (Fig. 15.4), horseshoe kidney (Fig. 15.5) are well seen on MRI.

15.1.4 Multicystic Dysplastic Kidney

Definition The kidney is replaced by noncommunicating cysts of varying sizes. It is due to atresia of the ureter or renal pelvis.

Imaging Features

• MRI is useful in complex/bilateral cases.

15.1 Genito Urinary System Anomalies



Fig. 15.4 Sonographic (a) image shows an enlarged kidney in the right renal fossa (arrow). Sagittal T2w MR image (b) shows crossed fused ectopia of the left kidney (open arrows)



Fig. 15.5 Axial T2W image showing horseshoe kidney (arrows)

- The affected kidney shows the absence of reniform shape. Noncommunicating peripheral and central cysts seen replacing the kidney (Fig. 15.6).
- The lack of normal renal parenchyma results in hypointensity on diffusion-weighted images.
- Bilateral renal involvement may lead to anhydramnios and pulmonary hypoplasia.

Differential Diagnosis

Lymphangioma—multi-septated cyst showing infiltrative features.

Enteric/Duodenal duplication cyst—may resemble perinephric fluid collection, shows laminated bowel signature.

Urinoma—Urine collection within Gerota's fascia due to leakage of urine from severe hydronephrosis. Seen as unilocular cyst that displaces the kidney.

Hydronephrosis due to pelvi-ureteric junction obstruction—The centrally placed dilated renal pelvis shows intercommunication with the peripherally placed dilated calyces.

Mesenteric cyst, Meconium pseudocyst are the other differential diagnoses.

15.1.5 Obstructive Uropathy

The common causes of fetal urinary tract dilatation are Pelviureteric junction obstruction, posterior urethral valves (PUV), ureterocele (usually associated with a duplication anomaly), vesicoureteric reflux disease, urethral atresia, and megacystis microcolon hypoperistalsis syndrome.

Oligohydramnios and pelvic bone ossification may obscure details on sonography. MRI is useful to identify perineal structures like the rectum, vagina, and uterus besides the features like over distended bladder, dilated ureters, and dilated pelvicalyceal system on MRI. Rectum, large bowel appear hyperintense on T1WI due to meconium.



Fig. 15.6 Sonographic (a) and axial MR T2W (b) images of fetal abdomen show an enlarged kidney with multiple non-communicating cysts suggesting multicystic dysplastic kidney (arrows)



Fig. 15.7 Sonographic (**a**) and axial MR T2W (**b**) images of fetal abdomen show horseshoe kidney (arrowhead). Hydronephrosis with pelvic ureteric junction obstruction is seen in the right limb (arrows). The bridging tissue could not be appreciated on USG

Cloacal abnormalities, posterior urethral dilatation, and ectopic ureteral insertional anomalies are better diagnosed by MRI [6, 7]. MRI can also be utilized for providing additional useful information on other renal features like cortical thickness and signal intensity of renal parenchyma. MRI is also useful to demonstrate cortical thinning in severe hydronephrosis. Dysplastic changes in the kidney lead to increased parenchymal T2 hyperintensity.

Pelvi-Ureteric Junction Obstruction

- Hydronephrosis with narrowing at Pelvi-ureteric junction (Fig. 15.7).
- Presence of communicating cysts within the kidney [7].

Prognosis

Prognosis depends on the contralateral kidney; milder anomaly being associated with a good prognosis. Vesico ureteric reflux, obstruction can be present in the contralateral kidney. Nonfunctioning kidney may involute postnatally.

Posterior Urethral Valve

- Bilateral ureters and pelvicalyceal system may be dilated (Fig. 15.8a).
- Dilated posterior urethra is seen as a "Keyhole." Urinary bladder appears distended (Fig. 15.8b).
- MRI is also useful to demonstrate cortical thinning in severe hydronephrosis.



Fig. 15.8 Axial (**a**) and sagittal (**b**) T2 W images of a fetal abdomen showing bilateral hydronephrosis (arrow) and an overdistended bladder (open arrows). Postnatal MCU (**c**) shows dilated posterior urethra (bro-

ken arrows), thin stream in anterior urethra (arrowhead) confirming the diagnosis of posterior urethral valve

• There may be urinary ascites when there is a leak from the distended urinary tract (Fig. 15.9).

Differential Diagnosis

Prune belly syndrome—There is a triad of large hypotonic bladder, deficiency of the abdominal wall musculature with anterior abdominal wall distension and bilateral cryptorchidism (Fig. 15.10).

15.1.6 Mesoblastic Nephroma

Definition Hamartoma of fetal kidney. It is a benign spindle cell containing tumor arising from the renal mesenchyme.

Imaging Features [8, 9]

Sonography: Solid mass with increased vascularity. Associated hypoechoic "ring sign" seen surrounding the tumor. Hydrops may be present.

MRI—Is useful to identify the organ of origin. The "Claw sign" is often seen suggesting intrarenal origin. The lesion is mildly hyperintense on T2WI. Larger masses may displace the bowel loops.

Differential diagnosis: Crossed fused ectopia, duplex collecting system, adrenal mass.

Prognosis: Better to be monitored in tertiary care hospital. Postnatal resection with margins is curative. Local recurrence or distant metastasis is rare.

15.1.7 Adrenal Neuroblastoma

Definition Malignant tumor comprising of neuroblasts, arises from adrenal medulla or sympathetic chain.

Pathology Normal fetal adrenal has neuroblastic nodules similar to neuroblastoma. These nodules involute normally. When they do not involute and keep growing, they evolve into neuroblastoma.

Staging:

Stage 1: Confined to the adrenal gland.

Stage 2: Showing extra-adrenal extension but does not cross midline.

Stage 3: There is extension across midline.

Stage 4: Distant metastases.

Stage 4S (Special): Shows good prognosis with metastases to liver, skin, bone marrow, and not bone.

Imaging Features

Sonography: Solid echogenic mass or complex cystic mass with thick septae and doppler flow. Hydrops may be present [10-12].

MRI—Useful to identify the organ of origin and to assess locoregional spread.

Solid mass—Hypo on T1 and moderately hyperintense on T2WI.

Cystic mass—Hypo on T1 and very hyperintense on T2WI.

Differential Diagnosis

- 1. Extra-lobar sequestration—common on left side (90%). It appears as a homogenous hyperintense mass on T2WI with a prominent feeding vessel from the aorta. A normal adrenal gland may be seen separately.
- 2. Adrenal hemorrhage: Very rare, showing blooming on GRE/EPI GRE images. By the signal characteristics, it is possible to identify the stage of hemorrhage.
- 3. Duplex Collecting system: The hydronephrotic upper moiety may resemble a cystic suprarenal mass. The



Fig. 15.9 Axial (**a**, **b**) and sagittal (**c**) T2 W images of a fetal abdomen showing bilateral hydronephrosis (arrows), overdistended bladder (open arrows), and urinary ascites (broken arrows)

hydronephrotic upper moiety may be associated with ectopic ureterocele. MRI is useful to identify a normal adrenal gland separately.

Prognosis Is variable with many of them remaining stable and some them showing resolution. Few of them can develop hydrops, extensive disease, and death.

Treatment

Postnatally stable masses less than 5cm size are followed up. Surgery is performed when masses are larger than 5 cm.

Chemotherapy is the option when there is a widespread disease.

15.1.8 Ovarian Cyst

Definition Commonly a benign cyst arising from the ovary.

Imaging Abdominal/pelvic cyst with imperceptible wall, with or without daughter cyst (Fig. 15.11) and may show attachment to the ovary. When there is torsion, the cyst may



Fig. 15.10 MR (a) T2W image of a 17-week fetus shows a large bladder (arrow) due to Prune belly syndrome, that was confirmed postnatally (b)



Fig. 15.11 Sonographic (a) and sagittal MR T2W (b) images of fetal abdomen show an abdominopelvic cyst on left side (arrow)—ovarian cyst. The cyst resolved on postnatal follow-up US scans

show fluid–fluid level and debris with associated free fluid in the abdomen [13, 14].

MRI is useful to identify the anatomy, complexity, and details like intracystic hemorrhage.



Fig. 15.12 T2W image in a 22-week fetus shows normal lower limb (arrow). The foot is seen at right angle to leg and is seen in profile

Differential Diagnosis Urachal cyst—midline cyst between bladder and cord insertion site.

Enteric duplication cyst—Presents in II trimester and shows "gut signature".

Mesenteric cyst—very rare and can resemble ovarian cyst, may be seen in midline [14].

Hydrocolpos—Midline and is seen posterior to bladder.

Prognosis Cysts less than 5 cm in size may resolve spontaneously. Cysts larger than 5 cm in size are unlikely to resolve and are prone to torsion. They can be treated by in utero- or postnatal aspiration.

Complication Torsion, Infarction/hemorrhage within the cyst can occur. They rarely cause intestinal obstruction because of the mass effect on the bowel [13, 15].

15.2 Musculoskeletal System

Ultrasonography, especially with three-dimensional imaging is preferred to study the Musculoskeletal system because of rapid limb movements. Apart from the usual HASTE sequence (Figs. 15.12 and 15.13) Echoplanar imaging (EPI), thick slab T2 sequences, and dynamic sequences can provide information about the skeleton and muscles. EPI is also useful to obtain an overview of the fetal thorax and skeleton.

Congenital Talipes Equinovarus

Normally in sagittal sections of the leg, the foot is seen at right angle and is seen in profile. In talipes equinovarus, in



Fig. 15.13 T2W images (a-c) in a 22-week fetus show normal upper limb (arrows) and hands (open arrows)



Fig. 15.14 T2W image in a 23-week fetus shows talipes of the foot (arrow). Though the leg is seen in profile (sagittal) view, the foot is seen enface

sagittal sections of the leg—the foot is seen medially deviated and is seen enface (Fig. 15.14).

15.3 Miscellaneous Fetal Conditions

15.3.1 Fetal Growth Restriction (FGR)

Synonym Intrauterine growth restriction (IUGR).

Definition and Pathology

It is defined as a condition where the estimated fetal weight (EFW) is less than the 10th percentile for the gestational age. There are two types:

- 1. Asymmetric FGR: The abdominal circumference is more reduced as compared to head circumference.
- 2. Symmetric FGR: The fetus is uniformly small and shows a reduction of all the relevant biometry values.

It is associated with several conditions—Chromosomal disorders, Several syndromes, congenital anomalies, Infection and Multiple gestations.

Imaging Features

Sonography with Doppler is the modality of choice to detect FGR. A knowledge of MRI in FGR is needed as it may be associated to a primary condition like Infection.

• MRI is very useful to calculate volumes of brain, body, lungs, and Placenta

- Placental volume/thickness may be reduced.
- Volumes of brain, body, lungs may be reduced. The usual parameters like BPD, HC, AC are also reduced.
- ADC values of some parts of the brain like Frontal white matter, centrum semiovale, thalami may be reduced [16].
- Placental perfusion is reduced [17].

15.3.2 Hydrops Fetalis

Definition and Pathology

Defined as accumulation of fluid in fluid two or more body cavities.

It is considered as a form of cardiac failure in fetuses. There are two types:

Immune hydrops fetalis—It occurs when there is fetomateral blood group/rhesus incompatibility.

Non-immune hydrops fetalis. Several causes have been identified to cause non-immune hydrops [18–20].

- 1. Cardiac anomalies, arrhythmia
- Chromosomal anomalies—trisomy 13,18,21, Turner syndrome
- 3. Infections
- 4. Pulmonary anomalies
- 5. Certain skeletal dysplasias
- 6. High output flow states/AV shunts
- 7. Twin-twin transfusion syndrome
- 8. Metabolic disorders
- 9. Fetal masses

Imaging Features

- Fetal anasarca
- Skin/subcutaneous/Scalp edema (skin thickness > 5 mm) (Fig. 15.15)
- Ascites/Pleural effusion/Pericardial effusion
- Placentomegaly (placenta thickness > 40 mm in II trimester, >60 mm in III trimester)
- Hepatosplenomegaly
- · Polyhydramnios

Prognosis

The prognosis is variable and is dependent on the causative factor.

15.3.3 Twin–Twin Transfusion Syndrome (TTTS)

Definition and Pathology

It is a prenatal condition in which monochorionic twins obtain unequal volume of placenta's blood supply. This results in the two fetuses growing at different rates.



Fig. 15.15 T2W image of a twin shows diffuse subcutaneous edema (arrow) due to hydrops

In TTTS there are abnormal vascular (arteriovenous and arterioarterial) anastomoses in the placenta. Hence, the placental circulation is directed predominantly toward one twin and is supplied less to the other twin [21]. The resultant hypoperfusion in one twin leads to oliguria in the hypovolemic (donor) twin with consequent oligohydramnios. There is hypervolemia and hypertension in the other (recipient) twin with consequent polyhydramnios.

Demise of one of the fetuses in a monochorionic pregnancy is associated with a risk of hypotensive ischemic injury in the surviving twin due to the "twin embolization" syndrome. As vascular anastomoses exist between twins due to monochorionic placentation—Demise of one twin leads to sudden loss of placental bed vascular resistance. The live twin becomes acutely hypotensive resulting in ischemic lesions of brain and kidneys.

Imaging Features [22] Recipient Twin

- Polyhydramnios
- Large urinary bladder
- Features of cardiac overload—cardiomegaly/fetal hydrops

Donor Twin

- Oligohydramnios, which may result in the twin appearing "stuck" to the side of the gestational sac.
- · Small or non-visualized urinary bladder
- Though MRI is not commonly used in the initial evaluation of TTTS, it is very useful in the identification of cerebral ischemia and intraventricular hemorrhage associated with TTTS.
- There may be altered hemodynamics leading to dilated cerebral venous sinuses in donor and recipient twins which may be well demonstrated on MRI.
- There may be dilatation of the renal pelvic collecting system in the recipient twin.
- Fetal MRI is useful after laser therapy for TTTS or after death of one twin to rule out ischemic (Fig. 15.16)/hemorrhagic injury to the brain parenchyma. There can be secondary ventriculomegaly secondary to ischemic encephalomalacic changes in the brain (Fig. 15.17).

Laser coagulation is the treatment of choice for TTTS in pregnancies at <26 weeks—Selective coagulation of the anastomoses at placental vascular equator is commonly performed.

- DWI can demonstrate cerebral damage immediately after fetoscopic laser coagulation if done 1–4 days after the procedure [23].
- MRI at 4–6 weeks after the procedure is useful to see brain changes like encephalomalacia.

15.3.4 Single Umbilical Artery

It is an important marker for chromosomal anomalies. It is the result of agenesis or atrophy of one of the arteries. The umbilical cord normally has two arteries and one vein (Fig. 15.18b). Single umbilical artery should always be ruled out during a routine second-trimester scan and can be detected on cross-sectional imaging of the umbilical cord (Fig. 15.18a).

It can be associated with trisomy 18, 13 and triploidy, fetal growth retardation, abnormalities of cardiovascular, genitourinary, gastrointestinal, skeletal, and central nervous systems.



Fig. 15.16 Axial diffusion-weighted images (a, b) showing acute infarcts (arrows) in bilateral parietal region in the surviving twin following cotwin demise. ADC images (c, d) showing restricted diffusion (open arrows)



Fig. 15.17 Sonographic (a) and MR (b, c) T2W images show bilateral ventriculomegaly (arrows) due to encephalomalacic changes in bilateral parieto-occipital lobes (open arrows) in the surviving twin following co-twin demise at 23 weeks. The present scan was taken at 29 weeks



Fig. 15.18 T2W image in a 24-week fetus (a) shows single umbilical artery (arrow); in a 31-week fetus (b) 2 umbilical arteries are seen (open arrows)

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