Imaging of cystic abdominal and pelvic masses in infants

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Authors: S. Gupta, P. Gahlot, J. Kapur; Singapore/SG
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Learning objectives

- The aim of this exhibit is to briefly describe the clinical presentation, pathology, radiological features and management of commonly occurring cystic abdominal and pelvic masses in infants.
- Pathologies have been categorized based on the organ of involvement.

Background

- Cystic masses are frequently encountered in infancy.
- These can be congenital or acquired.
- Imaging together with clinical findings can provide a reasonable differential diagnosis and guide further management, also helping to identify the patient's requiring surgical management.
- Here, we review the imaging features of the commonly seen abdominal and pelvic cystic masses in infants.

Findings and procedure details

**HEPATOPANCREATEOBLIARY SYSTEM**

**Hepatic cysts**

- Simple hepatic cysts are true cysts lined with cuboidal epithelium.
- Can be congenital or acquired (secondary to infection or trauma).
- These are often asymptomatic; rarely, symptoms may occur due to mass effect from large size.
- On ultrasound (US) a simple hepatic cyst is seen as anechoic lesion with an imperceptible wall and posterior acoustic enhancement.
- On CT and MRI, shows a well-defined lesion nearing water density and intensity, respectively with an imperceptible wall. These do not enhance.
- Complex cysts demonstrate internal septation in addition to the above findings.
- No further follow up is recommended for these lesions.

Hepatic mesenchymal hamartoma (HMH) [Fig. 1 on page 8] [Fig. 2 on page 9] [Fig. 3 on page 10]
• These are composed of bland spindle cells in myxoid to fibrous stroma with pseudocysts and normal appearing bile ducts and strands of hepatocytes.
• HMH can be multilocular cystic, cystic with solid nodule or less commonly predominantly solid.
• Patients present with non-tender enlarging abdominal mass, ascites, jaundice or congestive heart failure.
• On US, these are seen as anechoic multilocular cystic mass with thin walls sometimes with a solid nodule. Rarely, these are predominantly solid.
• On CECT the density of the lesion varies from near water density to high density fluid depending of protein content. The enhancement of the wall, septa and solid nodule depends on the stromal component.
• On MRI the lesions are hyperintense on T2W while the T1W signal can be hypointense or hyperintense depending on the water and protein content; enhancement pattern is similar to CECT scan.
• HMH is known to resolve with medical therapy although surgical resection is often required in larger lesions.

Hepatic abscess: pyogenic, amoebic or fungal

• Usually seen in premature newborns or immunocompromised infants secondary to systemic sepsis, biliary infection, trauma or ischaemia of liver or infection in the portal drainage area.
• Risk factors predisposing neonates to abscesses include umbilical catheterisation, long-term parenteral nutrition, prematurity and necrotising enterocolitis.
• Patients present with septicaemia and right upper quadrant tenderness.
• On US, these are seen as solitary or multiple thick walled hypoechoic unilocular or multilocular/septated cysts with increased surrounding vascularity.
• CECT shows a low-density lesion with or without internal septation and a thick enhancing rim and surrounding oedema.
• On MRI, these are T2W hyperintense, T1W hypointense with restricted diffusion on DWI/ADC. These have thick enhancing rim and surrounding inflammatory oedema.
• Drainage and aspiration is done under ultrasound or CT guidance followed by systemic antibiotic therapy, based on the microbiological culture and sensitivity of the aspirate.

Choledochal cysts (CCs) Fig. 5 on page 12

• CCs occur due to embryonic malformation of pancreato-biliary ductal union.

Todani classification: Fig. 4 on page 11

• Type 1: Segmental and diffuse dilatation of the common bile duct
• Type 2: Diverticulum of extra-hepatic duct
• Type 3: Choledochocele
• Type 4: (a): Multiple cystic dilatations of extra and intrahepatic bile ducts
• Type 4: (b): Multiple cystic dilatation of the extrahepatic bile duct
• Type 5: Multiple cystic dilatations of the intrahepatic bile ducts

• Patients present with abdominal pain, jaundice and RUQ mass.
• On US these are seen as a cystic structure in the peripancreatic region communicating with the biliary tree, separate from the gallbladder.
• MRCP demonstrates their communication with biliary ducts, relations to surrounding structures and also helps in classifying the type.
• These cysts are surgically resected and many investigators believe Roux-en-Y hepatico-jejunostomy to be the best operative approach.

**Pancreatic cysts**

• Pancreatic cysts are true cysts lined with flattened glandular epithelial lining.
• These are developmental anomalies due to sequestration of primitive pancreatic ducts; these do not contain pancreatic enzymes.
• Can be associated with various syndromes such as autosomal dominant polycystic kidney disease and tuberous sclerosis.
• Presenting symptoms include abdominal distension, vomiting, jaundice or pancreatitis.
• On US, these are seen as unilocular or multilocular anechoic cystic lesions in the body or tail of the pancreas demonstrating posterior acoustic enhancement.
• MRI compliments the ultrasound findings and shows water intensity in cysts on T1W and T2W images. MRI is better at demonstrating the relationship with surrounding structures.

**Pseudocysts** Fig. 6 on page 13

• The walls of pseudocysts are lined with fibrous tissue.
• These cysts contain pancreatic enzymes as opposed to the true pancreatic cysts.
• These are formed as a result of single or repeated episodes of pancreatitis.
• On US, these are seen as anechoic cysts with posterior acoustic enhancement in the epigastrium, usually related the pancreas; internal echogenicity can be seen due to debris.
• CT or MRI may be needed for treatment/surgical planning. In addition to the above findings, CT/MR can show signs of recurrent or chronic pancreatitis such as calcification and dilated ducts.
• Treatment includes expectant management, internal or external drainage.
• Analysis of the drained fluid is often helpful in confirming the diagnosis.

**Kidneys**

**Multilocular cystic renal tumors** Fig. 7 on page 14 Fig. 8 on page 15
• These are a spectrum of cystic renal lesions ranging from cystic nephroma (lined by epithelium and fibrous septa with mature tubules) and cystic partially differentiated nephroblastoma (the septa have blastemal cells).
• Patients usually present with painless abdominal mass.
• Imaging cannot distinguish between the two types of lesions.
• On ultrasound these lesions appear as an encapsulated mass with anechoic cysts of varying size confined to or extending through the renal capsule.
• Differentials include cystic Wilms tumour (has a solid expanding nodule).
• Nephrectomy is the treatment of choice and patients have an excellent prognosis in complete tumour excision.

**Autosomal recessive polycystic kidney disease (ARPKD)**

• Depending on the age of presentation ARPKD is subcategorised as: perineonatal, neonatal, infantile and juvenile.
• The disease occurs as a result of mutation of PKHD1 gene, which encodes fibrocystin, a protein.
• On US multiple anechoic cysts of increasing size are seen in both kidneys and liver.
• CT and MRI compliment the findings of ultrasound.
• With time kidneys lose their concentration ability and eventually systemic hypertension develops.
• Hepatic fibrosis associated with ARPKD develops in childhood or may rarely be congenital.
• Perineonatal and infantile subgroups have the worst prognosis.

**Multicystic dysplastic kidneys (MCDK)**

• MCDK is characterised by presence of islands of undifferentiated mesenchyme, often with cartilage and immature collecting system.
• The cysts can vary in size from microscopic to over several centimetres in one or both the kidneys.
• On US, these are seen as multiple cystic lesions in the kidneys not communicating with collecting system; no renal tissue is identified.
• MR Urography is better at demonstrating lack of communication with the collecting system.
• DMSA scan does not show radiotracer uptake in the kidneys due to absence of functioning tissue.
• There is no evidence that MCDK is a premalignant condition, nephrectomy is therefore not necessary in asymptomatic cases.
• Every 6 monthly follow up with imaging is recommended for 3 years from diagnosis.
• If cysts increase in size nephrectomy is performed to rule out underlying Wilms tumour.
• In neonates with bilateral MCDK renal transplant is required for survival.
Hydronephrosis Fig. 13 on page 20 Fig. 14 on page 21 Fig. 15 on page 23

- Can be secondary to ureteropelvic junction obstruction (UPJO), vesicoureteric reflux or posterior urethral valves.
- In addition congenital megaureters can also present as cystic abdominopelvic mass.
- UPJO is the most common cause of antenatal hydronephrosis.
- It is anatomical or functional obstruction leading to an aperistaltic segment resulting in partial or complete obstruction at ureteropelvic junction.
- US shows gross dilatation of the collecting system with transition to normal caliber distal to the ureteropelvic junction.
- An antegrade nephrostogram shows hydronephrotic kidney with transition to normal caliber at ureteropelvic junction.
- Percutaneous nephrostomy can be performed as first step prior to definite surgery (Pyeloplasty) to decompress the system.

Bowel and mesentery

Lymphangioma Fig. 16 on page 23 Fig. 17 on page 24

- These are lesions of vascular origin that show lymphatic differentiation.
- 95% of these lesions occur in the neck and axillary region, in the abdomen, the most common location is mesentery.
- Lymphangiomas are large multilocular cystic lesions that may show dependent debris and internal haemorrhage.
- On US these are large multilocular anechoic cysts that may demonstrate internal echogenicity reflecting internal haemorrhage or debris.
- MRI and CT scan compliment the findings on ultrasound and are often required in large Lymphangioma for surgical planning.
- Surgical excision is the treatment of choice.

Enteric duplication cysts Fig. 18 on page 25

- These occur anywhere along the alimentary tract.
- Histologically these resemble bowel loops with internal mucosa and outer muscular layer.
- US and barium studies are often sufficient for diagnosis, MRI and CT scan being reserved for difficult cases.
- US reflect the histological features with echogenic internal wall and hypoechoic outer wall described in literature as "Double Wall Sign".
- On CT these cysts have a lobulated appearance and are hypodense to hyperdense depending on contents.
- On MRI, these are hyperintense on T2W images and hypointense to hyperintense on T1W depending on the protein content.
- Differentials on imaging include intussusception and appendicitis.
• Malignancy associated with these cysts is rare and are usually seen in the adult population.
• Surgical resection is the treatment of choice.

**Dilated bowel loops** [Fig. 20 on page 27 Fig. 21 on page 29]

• Can be seen secondary to obstruction due to various causes at various levels and can present as cystic abdominal masses.
• The obstruction can be at the level of:
  • Pylorus in hypertrophic pyloric stenosis (HPS).
  • Proximal small bowel due to congenital bands or malrotation.
  • Ileocolic junction due to intussusception.
  • Distal colon due to Hirschprung disease.

**Pelvic cavity**

**Hematocolpos or hematometrocolpos** [Fig. 22 on page 29]

• These are distension of the vaginal cavity and vaginal cavity and the uterus, respectively.
• These occur secondary to vaginal outflow obstruction often due to vaginal stenosis or imperforate hymen.
• Infants usually present with midline abdominal mass, abdominal distension and urinary obstruction.
• On US there is a midline anechoic cystic lesion in the pelvis. Internal echoes when seen represent mucoid material and cellular debris. Fluid-Fluid level is seen if the contents are hemorrhagic.
• MRI signal intensity on T1W and T2W sequences depends on the content of the lesion and presence or absence of haemorrhage.
• The imperforate hymen or vaginal stenosis is surgically treated and prognosis is good after successful surgical therapy.

**Ovarian follicles**

• These are small thin walled anechoic ovarian cysts.
• Generally seen in post-pubertal girls but can also be seen in neonates and infants due to elevated levels of maternal hormones.
• These are often asymptomatic are are brought to medical attention due to complications of haemorrhage or rupture.
• On US, these are seen as thin walled cystic masses, no further imaging is usually needed.
• Follow up with ultrasound until complete resolution is however suggested to demonstrate true physiological nature.
Mature cystic teratoma (MCT) Fig. 23 on page 30

- MCT are benign adnexal lesions that consist of tissue from at least two of the three germ layers (endoderm, mesoderm or ectoderm).
- Sacrococcygeal area is the most common site (47.2%) followed by gonads (31.6%).
- Clinically infants present with a midline mass or lower abdominal pain and tenderness when torsion sets in.
- Plain films can demonstrate calcifications resembling a tooth.
- US demonstrate hypoechoic fluid content with hypoechoic and hyperechoic solid component with posterior acoustic shadowing from the ectodermal component.
- The T1W and T2W signal of the lesion is dependent on tissue content; fat is bright on both T1W and T2W images while calcium is seen as signal void on both. The solid component also demonstrates enhancement post contrast administration.
- Upto 15% of teratomas are associated with ovarian torsion.
- Surgical resection is performed because of risk of malignancy.

Images for this section:
**Fig. 1:** Hepatic Mesenchymal Hamartoma in an 11 month old female who presented with abdominal distension for 3 months. US shows multiloculated cystic lesion occupying a greater part of the liver, mainly the right lobe.
Fig. 2: Same patient as Fig 1. CE T1-W image shows large cystic lesion in the liver with thin enhancing septation.
Fig. 3: Same patient as in Fig 1 and 2. coronal T2-W image shows large cystic lesion in the right hepatic lobe with thin internal septation.
**Fig. 4:** Todani classification of Choledochal cysts, a schematic drawing.
**Fig. 5:** Newborn male with a RUQ mass noted on routine post-natal physical examination. US shows single fusiform dilatation of the common duct in keeping with Type 1 Choledochal cyst; the gallbladder was separately seen. The child subsequently underwent resection of the cyst with Roux-en-Y hepaticojejunostomy.
**Fig. 6:** A 6 month old male child operated before for Choledochal cyst: Transverse US image in the upper midline abdomen shows anechoic peripancreatic fluid collection thought to represent a pseudocyst. Ultrasound guided external drainage was performed with subsequent resolution of cyst.
Fig. 7: Transverse US image of the right kidney of a 12 month old female child showing multiloculated cystic lesion in keeping with cystic nephroma. The cyst remained stable on follow up US until 5 years. On subsequent US at 12 years (not shown) there was increase in size. A PET CT was performed- Fig 8
**Fig. 8**: PET CT of the same child (Fig.7) performed at 12 years of age, shows no increase in uptake of radiotracer.
Fig. 9: US image of a newborn male showing multiple cystic lesions in the right renal bed. No renal tissue was identified. Findings suggest multi-cystic dysplastic kidney
Fig. 10: Coronal MR Urogram image of the same child (Fig.9) shows multiple cystic lesions in the right renal bed with no communication with the collecting system. Tortuous right ureter is noted with ectopic insertion in the urethra (not shown). Incidental left ureterocele is noted.
**Fig. 11:** DMSA of the same child (Fig.9 and 10) shows no uptake of radiotracer in the right kidney suggesting no functional renal tissue.
**Fig. 12:** US image of the right renal bed in the same child (Fig.8, 9, and 10) at 4 years of age shows no identifiable renal tissue; in addition, a single sub-centimeter cyst was seen and all the other previously seen cysts had resolved.
Fig. 13: Ballotable left kidney noted in routine post-natal examination. Longitudinal US image of the left kidney shows gross left sided hydronephrosis. The left ureter was not seen.
**Fig. 14:** Post-void MCU image of the same child (Fig. 13) does not show any reflux of contrast into the ureters or renal pelvis.

**Fig. 15:** Antegrade nephrostogram performed while placement of PCN tube shows gross left hydronephrosis with transition to normal size left ureter at the uretero-pelvic junction. Findings suggest left urtero-pelvic junction obstruction.
**Fig. 16:** Antenatal US on this neonate showed ascites (not shown). On post natal US a large anechoic cystic mass with septations was seen in the abdomen.
Fig. 17: Axial CT image of the same child (Fig.16) shows multiloulated cystic lesion in the left anterior abdominal wall extending in to the peritoneal and left retroperitoneal cavity; the left kidney is displaced anteriorly. Findings are in keeping with lymphatic malformation. The lesion was surgically resected and diagnosis confirmed on histology.
Fig. 18: Intra-abdominal cystic lesion was detected in antenatal US at 24 weeks (not shown). In the post natal US image a cystic lesion was seen with inner hyperechoic and outer hypoechoic rim; a peripheral soft tissue nodule was noted. Working differentials were ovarian cyst, enteric duplication cyst and mesenteric cyst
**Fig. 19:** Contrast enhanced T1-W F/S MR image showing a large cystic lesion anterior to the left kidney corresponding to the cyst seen on US (Fig.18). During surgical resection the origin was noted approximately 5 cm distal to the duodeno-jejunal junction. Histology confirmed enteric duplication cyst.
**Fig. 20:** An abdominal radiograph of a new born male child taken after instilling contrast through the indwelling nasogastric tube shows gross dilatation of the proximal small bowel. Findings were in keeping with proximal small bowel obstruction.

**Fig. 21:** US image of the left upper quadrant of the same child (Fig.19) shows grossly distended jejunum. no intussusception or cause for obstruction was identified. During surgery a congenital band was noted across the proximal small bowel causing obstruction and bowel wall ischemia.
Fig. 22: A 9 month old child with absent right kidney noted at birth (not shown) presented with lower abdominal distension. Longitudinal US image of pelvis showing distension of the uterus with anechoic fluid in keeping with hydrometra. The bladder is compressed anteriorly.
**Fig. 23:** 9 month old child with known right ovarian teratoma at birth (image not shown) for expectant management presented with sudden onset lower abdominal pain. US image of the pelvis shows an echogenic lesion with no posterior acoustic shadowing (fatty content) with a large anechoic cystic component. Right ovarian torsion was noted at surgery and histology suggested mature cystic teratoma with ovarian torsion.
Conclusion

- Imaging together with a good clinical history and physical examination findings can often provide a reasonable differential diagnosis.
- This is helpful in determining the follow up plan and differentiating surgical from the non-surgical cases.

Personal information

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