Abdominal fluid containing masses of the neonatal period: all you need to know

Poster No.: C-1499
Congress: ECR 2013
Type: Educational Exhibit
Authors: C. Bruno, G. Caliari, N. Zampieri, R. Pozzi Mucelli; Verona/IT
Keywords: Paediatric, Abdomen, Obstetrics (Pregnancy / birth / postnatal period), Ultrasound, MR, Conventional radiography, Barium enema, Intrauterine diagnosis, Comparative studies, Congenital, Obstetrics
DOI: 10.1594/ecr2013/C-1499

Any information contained in this pdf file is automatically generated from digital material submitted to EPOS by third parties in the form of scientific presentations. References to any names, marks, products, or services of third parties or hypertext links to third-party sites or information are provided solely as a convenience to you and do not in any way constitute or imply ECR’s endorsement, sponsorship or recommendation of the third party, information, product or service. ECR is not responsible for the content of these pages and does not make any representations regarding the content or accuracy of material in this file.

As per copyright regulations, any unauthorised use of the material or parts thereof as well as commercial reproduction or multiple distribution by any traditional or electronically based reproduction/publication method is strictly prohibited.

You agree to defend, indemnify, and hold ECR harmless from and against any and all claims, damages, costs, and expenses, including attorneys’ fees, arising from or related to your use of these pages.

Please note: Links to movies, ppt slideshows and any other multimedia files are not available in the pdf version of presentations.

www.myESR.org
Learning objectives

To offer a complete overview of the diagnostic hypotheses of the various fluid-containing masses which can be found in the neonatal abdomen in order to address the correct diagnostic pathway.
Background

The finding of a fluid-containing mass is not a rare event in the Imaging evaluation of the neonatal abdomen. Most of them are detected during intra-uterine life; sometimes they can be discovered after delivery, often because of obstruction symptoms they cause. Regardless of the organ they belong to, they can be classified considering their nature into newly-formed masses, dilation of pre-existing hollow structures or persistent embryological remnants.

A) Newly-formed masses. This is a miscellaneous group in which we include cystic lesions which are not a normal part of the organ they origin from, possibly because of an error in the normal embryological development (e.g. gastroenteric duplication cysts) or of an abnormal hormonal stimulation.

a) Duplication cysts are a rare entity whose origin is to be found in an error of embryological segmentation. They can occur everywhere in the bowel but they are most frequently found in the jejunum or in the ileum. As it can easily be understood considering the name, at US - which plays a pivotal role in the diagnosis - their features are very similar to the normal bowel wall architecture ("muscular-rim sign"), showing a typical three-layer bowel-like pattern: echoic serosa, hypoechoic muscular layer and echogenic mucosa (Fig. 1 on page 8 , a, b). Less often, five sonographic layers can be detected, reflecting superficial mucosa, deep mucosa, submucosa, muscularis propria and serosa. Some false-positive diagnoses of duplication cysts have been described in Literature, especially when other fluid-containing masses undergo complications like it happens in case of torsion of ovarian cysts where internal debris can mimic a multi-layered wall. On the other hand, duplication cysts may be underdiagnosed in case of superimposed infections, when the identification of the eroded inner layers is difficult.

b) Choledocal cysts are a broad spectrum of congenital anomalies of the biliary tree characterized by the dilation of extraepatic and intraepatic ducts. The current Todani’s classification identifies five types (Fig. 2 on page 8, a-f).

Todani’s type I, divided into three subtypes, is a dilatation of the common bile duct of various length, which - according to many researchers - can be caused by an aberrant insertion of the pancreatic duct inducing a reflux of pancreatic enzymes into the biliary tree and therefore a progressive weakening of the wall.

Todani’s type II - which is supposed to be caused by antenatal rupture of the common bile duct - is characterized by the presence of one or more diverticula of the common bile duct (Fig. 3 on page 9).
Todani’s III consists in a choledococele, i.e. the dilatation of the common bile duct in its intraduodenal portion. The pancreatic duct and the common bile duct empty in it. The causes are not well understood; it can be due to either a duodenal duplication in the ampullary region or it can be secondary to obstruction.

Todani's IV is characterized by multiple extrahepatic (IV A and B) and intrahepatic (IV A) dilation (Fig. 4 on page 10).

Todani's V is Caroli's disease, i.e. dilation of multiple intrahepatic bile ducts.

US can easily depict the correct origin of the cysts and their origin from the biliary tree, showing the markedly enlarged duct as compared to the adjacent normal segments. If a correct diagnosis is difficult to be achieved MRI can be used as well, using a MRCP protocol.

c) Renal cysts can be found most frequently as a developmental dysplasia (multicystic dysplastic kidney) which is related to early obstruction during intra-uterine life leading to the failure of the correct junction between the distal portion of the nephrons and the pyelo-calyceal system. According to many reports 20-50% of patients with multicystic dysplastic kidney have a concomitant renal abnormality dominating the Imaging features. The cortex is underdeveloped with multiple, randomly distributed cysts of variable size (Fig. 5 on page 11). Unless the underlying renal abnormality is bilateral, multicystic dysplastic kidney is monolateral, which helps its differentiation from the typically bilateral autosomal-dominant polycystic disease. The diagnosis can be made by means of US which allows a correct evaluation of the degree of cortico-medullary differentiation and, if present, of the underlying renal malformation. In selected cases a correct anatomical depiction can be obtained with MR urography (Fig. 6 on page 12).

d) Ovarian cysts are probably the result of maternal hormonal stimulation. Small cysts (i.e. smaller than 1 cm) are not a rare finding and do not need follow-up. Occasionally they can grow to a larger size and can become a potential cause of ovarian torsion. US is able to detect them and to evaluate their content, considering that the finding of intra-cystic debris can be a sign of internal hemorrhage due to torsion (Fig. 7 on page 13 , a-c).

B) Dilation of pre-existing hollow structures can be due to two different mechanisms: downstream obstacles or reflux.

a) The gastro-enteric tract can be obstructed only with the first mechanism; causes are different and depend on the segment considered. Generally speaking bowel obstruction can be divided into high obstruction (i.e. until the proximal jejunum), characterized by less
than three dilated loops, and low obstruction (i.e. distal to the proximal jejunum) where more than three dilated loops can be found.

a) Among **high obstruction** gastric, duodenal and ileal obstruction can be found. Stomach atresia is very rare and characterized by the "single bubble" sign, whereas in duodenal atresia the "double bubble" sign is detected. In both cases no air in the distal segments can be found and diagnosis can be made on plain radiographs (Fig. 8 on page 14). Duodenal stenosis can be related to malrotation or volvulus, to annular pancreas, to duodenal web; in all these cases the clue to diagnosis is the finding of the "double bubble sign" with distal gas. An upper gastrointestinal examination with oral administration of contrast material leads to the diagnosis.

b) Among **low obstruction** (characterized by diffusely dilated loops) three different mechanisms can be described leading to occlusion and dilation of bowel loops: atresia (secondary to ischemic damage) (Fig. 9 on page 15), meconium ileus and Hirschsprung disease. Contrast enema is the method of choice to evaluate distal bowel occlusion. In the case of atresia it is able to depict the atresic segment. Meconium ileus is related to the inspissation of abnormal meconium, leading to mechanical ileal occlusion. It is often related to cystic fibrosis. Contrast enema shows microcolon distal to the obstruction and multiple filling defects secondary to meconium plugs (Fig. 10 on page 16, a, b). It must be differentiated from functional immaturity of the colon which is a self-limiting condition.

Hirschsprung disease is determined by the arrest of migration of ganglionic cells and it is often a distal functional occlusion; the affected segment is generally located to the rectum-distal sigma (although "long segment diseases" have been described). The clue to diagnosis is the demonstration, using contrast enema, of the transition zone between a normal or variably narrowed aganglionic distal segment and a dilated proximal segment (Fig. 11 on page 17).

b) The **dilation of renal pelvis and ureter** can be due to both mechanisms described before. Obstruction can be found either at the pyelo-ureteral junction leading to important distension of the pelvis and calices without evidence of the ureter (Fig. 12 on page 18, a, b), or at the intravesical segment of the ureter (ureterocele). US is able to diagnose both conditions showing in the former a markedly dilated pelvis and in the latter the ureterocele appearing as a sonolucent cystic structure with a thin echoic wall (Fig. 13 on page 19, a, b).

Less frequent causes of low ureteral obstruction are bladder diverticula, often located at the vesico-ureteral junction (Hutch diverticulum). Besides US, MR urography can provide excellent demonstration of the disease, depicting also the possible related kidneys anomalies (i.e. ectopic ureter, pelvis duplication) (Fig. 14 on page 20, a, b).
Another cause leading to pyelo-ureteric dilatation is reflux which can be both primary or secondary. Differential diagnosis between low urinary tract obstruction and reflux can be achieved only using voiding cystourethrogram (Fig. 15 on page 21) since US and MR can only depict anatomy.

c) **Bladder overdistention** is secondary to obstructive mechanisms: the most common cause is posterior urethral valves.

They result from the formation of a thick valvelike membrane from tissue of wolffian ducts that courses obliquely from the verumontanum to the distal part of the prostatic urethra. The resulting obstruction leads to a bladder wall thickening with elongation of the posterior urethra and hypertrophy of the bladder neck which appears narrow in relation to the dilated urethra. Secondary ureteral and pelvis dilation follows (Fig. 16 on page 21).

d) **Vaginal dilation** is due to obstructive mechanisms (Fig. 17 on page 22, a-c).

Imperforate hymen is the simplest cause of vaginal obstruction. The vagina is obliterated by a thin membrane which forms at the junction between the mullerian ducts and the urogenital sinus.

In case of a transverse vaginal septum the vagina is obliterated by fibrous connective tissue with vascular and muscular elements lined by squamous epithelium. The area can be a thin membrane but it can involve a segment of the vagina leading to segmental vaginal atresia.

The diagnosis can be made with US, showing the markedly distended vagina and the trasitional zone distal to obstruction (Fig. 18 on page 23). MRI can be used to depict more complex associated malformations as well as vaginogram or voiding cystourethrogram can show whether uro-genital fistulae are present.

C) A fluid-containing mass can be also the result of the **dilation of abnormal viscera** resulting from the persistence of embryonal structures.

The most common of them are cloacal malformation (Fig. 19 on page 24, a-d) and urogenital sinus persistence; they both derive from a failure of the urorectal septum in the division of the cloaca, which is the common channel where the embryonal urinary, gastrointestinal and genital tract empty during the first 5 weeks.

In the cloacal malformation the genital, the urinary and the gastrointestinal tract persist communicating and all drain through a common perineal opening; in the second one the urinary and genital tracts drain through a common opening and the gastrointestinal tract drains separately. US with transperineal approach can depict the exact communication of the three channels and evaluate kidneys for related abnormalities (Fig. 20 on page 25).
contrast enema and voiding cystourethrogram are essential for diagnosis (Fig. 21 on page 26 a, b) while MRI is useful to detect associated anomalies of the spinal cord.
Fig. 1: a, b: Duplication cists can occur everywhere in the gastrointestinal tract (a: duodenal duplication; b: ileal duplication). The characteristic aspect is the "muscular-rim sign", showing a typical three-layer bowel-like pattern: echoic serosa, hypoechoic muscular layer and echogenic mucosa (asterisk)

© Radiology, Policlinico GB Rossi - Verona/IT
**Fig. 2:** a-f: Todani's classification divides choledocal cysts into five subtypes with different embryological origin.

© Radiology, Policlinico GB Rossi - Verona/IT
**Fig. 3:** Todani’s type II cyst. US scan showing a large cyst close to the gallbladder.

© Radiology, Policlinico GB Rossi - Verona/IT
**Fig. 4:** Todani’s IV A malformation of the bile ducts. T2 weighted sequence with fat-saturation shows markedly dilated extrahepatic duct.

© Radiology, Policlinico GB Rossi - Verona/IT
Fig. 5: Multicystic displastic kidney is characterized by an underdeveloped cortex with multiple, randomly distributed cysts of variable size. Cortico-medullary differentiation is poor.

© Radiology, Policlinico GB Rossi - Verona/IT
**Fig. 6:** MR urography can help obtaining a complete anatomical depiction of underlying anomalies. Double ureter with markedly dilated upper pelvis and inferior megaureter with ectopic opening into bladder neck.

© Radiology, Policlinico GB Rossi - Verona/IT
**Fig. 7:** a-c: Simple cysts - which are probably the result of abnormal in utero stimulation - are not a rare finding (a). They can be easily evaluated by means of US which is able to depict internal debris (b, c) which are due to internal hemorrhage probably secondary to torsion.

© Radiology, Policlinico GB Rossi - Verona/IT
**Fig. 8:** Duodenal atresia is characterized by the "double bubble sign". No distal gas can be detected. (Courtesy of dr F.Moore).

© Radiology, Policlinico GB Rossi - Verona/IT
Fig. 9: Ileal atresia can be included among the low obstructions. It is caused in many cases by ischemic damage and is characterized by diffusely dilated loops. (Courtesy of dr F. Moore).

© Radiology, Policlinico GB Rossi - Verona/IT
Fig. 10: a, b: Meconium ileus is related to the inspissation of abnormal meconium. Contrast enema shows microcolon distal to the obstruction and multiple filling defects secondary to meconium plugs.

© Radiology, Policlinico GB Rossi - Verona/IT
**Fig. 11:** The clue to diagnosis for Hirschsprung disease is the demonstration, using contrast enema, of the transition zone (asterisk) between a normal or variably narrowed aganglionic distal segment and a dilated proximal segment.

© Radiology, Policlinico GB Rossi - Verona/IT
Fig. 12: a, b: among the causes leading to pelvis dilation obstruction at the pyelo-ureteral junction leading to important distension of the pelvis and calices without evidence of the ureter can be found. (a: longitudinal scan, b: axial scan)

© Radiology, Policlinico GB Rossi - Verona/IT
Fig. 13: a, b: US can depict ureterocele appearing as a sonolucent cystic structure with a thin echoic wall. On VCUG it appears with the characteristic "cobra head" aspect.

© Radiology, Policlinico GB Rossi - Verona/IT

Fig. 14: a, b: Less frequent causes of low ureteral obstruction are bladder diverticula, often located at the vesico-ureteral junction (Hutch diverticulum). They can be related to other kidneys' anomalies (i.e. ectopic ureter, pelvis duplication)

© Radiology, Policlinico GB Rossi - Verona/IT
**Fig. 15:** Passive high grade reflux. Voiding cystourethrogram is the clue to obtain diagnosis of vesicoureteral reflux. It can also differentiate between active (during voiding) and passive reflux.

© Radiology, Policlinico GB Rossi - Verona/IT
**Fig. 16:** Voiding cystourethrogram showing obstruction due to posterior urethral valves. Bladder wall is thickened with elongation of the posterior urethra and hypertrophy of the bladder neck which appears narrow in relation to the dilated urethra. Secondary ureteral and pelvis dilation follows.

© Radiology, Policlinico GB Rossi - Verona/IT
Fig. 17: a-c: Three different grades of vaginal obstruction can be found, ranging from atresia (a) to imperforate hymen (c), which is the simplest cause of vaginal obstruction. In case of a transverse vaginal septum the vagina is obliterated by fibrous connective tissue with vascular and muscular elements lined by squamous epithelium (b).

© Radiology, Policlinico GB Rossi - Verona/IT
Fig. 18: Transperineal approach. In the cause of vaginal obstruction diagnosis can be made by US which shows the markedly distended vagina (asterisk) and the transitional zone distal to obstruction.

© Radiology, Policlinico GB Rossi - Verona/IT
Fig. 19: a-d. In the cloacal malformation the genital, the urinary and the gastrointestinal tract persist communicating and all drain through a common perineal opening.

© Radiology, Policlinico GB Rossi - Verona/IT
**Fig. 20:** Urogenital sinus: US with transperineal approach can depict the exact communication of the urethra and the vagina (asterisk).

© Radiology, Policlinico GB Rossi - Verona/IT
Fig. 21: a, b: in cloacal malformation contrast enema and voiding cystourethrogram are essential for diagnosis, correctly depicting the common channel connecting the urethra, the vagina and the rectum (b: bladder; v: vagina; r: rectum)(courtesy of Dr F.Moore).

© Radiology, Policlinico GB Rossi - Verona/IT
Imaging findings OR Procedure details

Plain radiographs and transabdominal US are the first step and in some cases can lead to the correct diagnosis, In the evaluation of lower urinary tract or of complex genitourinary malformations other approaches such as the transperineal approach can give excellent details.

Contrast enema, upper gastrointestinal examination with oral contrast medium administration and voiding cystourethrogram still play an important role.

MRI with MRCP protocol or urography are mandatory when anatomical detail is needed.
Conclusion

The finding of a fluid-containing mass is not a rare event in the evaluation of the neonatal abdomen. General radiologists should be confident with the main imaging findings in order to choose the pathway leading to a correct diagnosis.


13) Ranganath SH, Lee EY, Eisenberg RL. Focal cystic abdominal masses in pediatric patients. AJR Am J Roentgenol. 2012; 199(1)
