The Y-configuration on pediatric ultrasound: yes, that's an enteric duplication cyst!

Poster No.: C-1806
Congress: ECR 2014
Type: Educational Exhibit
Authors: K. Kalogerakou, M. Vakaki, G. Karapostolakis, D. Grigoraki, C. Argiris, J. Koutsogiannis, O. Paskoviti, C. Koumanidou; Athens/GR
Keywords: Cysts, Congenital, Diagnostic procedure, Ultrasound, Paediatric, Gastrointestinal tract
DOI: 10.1594/ecr2014/C-1806

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 illustrate the importance of the Y-configuration sonographic sign for the definite preoperative diagnosis of an enteric duplication cyst in children.

- To familiarize pediatric radiologists with this sign, caused by the splitting of the shared muscularis propria layer between the cyst and the bowel, as a key sonographic finding.
**Background**

Enteric duplication cysts are uncommon congenital abnormalities that originate anywhere along the alimentary tract, from tongue to the anus. Most cysts are found in infants and children. The small bowel duplication cyst is the most common type of enteric duplication cyst and accounts for about 50% of all reported duplications, while the ileum is the most common location, followed by the esophagus, large bowel, jejunum, stomach and duodenum.\(^1,2,3\)

Duplications cysts occur on the mesenteric border of the small bowel and are either sausage-shaped, long and tubular or rounded structures or even mixed and they may or may not connect with the bowel lumen. The tubular duplications often communicate with the bowel and are more common in the lower gut and colon but the majority of duplications are noncommunicating spherical cysts of variable size. \(^2\) As the name suggests, an enteric duplication cyst, is histologically similar to the gastrointestinal tract that is lined with mucosa and submucosa surrounded by muscularis propria.\(^4\) Ectopic tissue can be found in the walls of these lesions at all levels of the gastrointestinal tract. Gastric mucosa and pancreatic tissue appear to be the only ectopic tissues found in the gastrointestinal tract duplications that are clinically significant. \(^3\) Malignancy, although rare, has also been reported in a female infant with a duplicated gut. \(^5\)

The majority of duplication cysts present during the first year of life, although some occasionally manifest in older patients. They may be silent and an incidental finding but when symptomatic, abdominal symptoms may include the presence of pain, masses and bleeding per rectum, in case of ectopic gastric mucosa, or complications such as intussusceptions, volvulus or intestinal obstruction. \(^3,5\)

Several theories have been proposed to explain the embryologic basis for gastrointestinal tract duplications. However, no single hypothesis can explain all possible combinations of duplications, locations and associated anomalies such as vertebral anomalies which can be a helpful clue in the radiographic diagnosis of duplications. \(^3\) The 'abortive twinning theory' proposes that gastrointestinal tract duplications represent incomplete twinning. This could explain the colorectal tubular duplications or duplications of the hindgut that are associated with duplication of genital and urinary structures. The 'aberrant luminal recanalization theory' is a popular theory that adequately explains duplications in those portions of the gastrointestinal tract that go through the 'solid stage', including the esophagus, small bowel and colon. However, this theory cannot be used for duplications at other levels. The 'persistent embryologic diverticula theory' suggests that small diverticula found on the antimesenteric aspect of the intestinal wall of embryos persist and develop into gastrointestinal tract duplications. The majority of the duplications however, are located on the mesenteric aspect of the gut, making this theory difficult to justify. The 'intrauterine vascular accident theory' suggests that like small bowel atresias, gastrointestinal duplications arise from an intrauterine vascular
accident during early fetal development and may be a valid explanation for small bowel atresias and perhaps their associated duplications. (2,3) The 'split notochord theory' is the accepted explanation for the so called neurenteric cysts, the enteric-lined cysts located in the posterior mediastinum, abdomen or spinal canal associated with adjacent vertebral anomalies. (3,5)

The most common modalities used to image duplications are ultrasound and contrast medium examinations. Barium studies usually reveal an intraluminal, intramural or extrinsic mass. Technetium-99m pertechnetate scintigraphy can be helpful for suspected esophageal, duodenal, and small-bowel lesions that contain ectopic gastric mucosa. CT and MRI are not the usual methods for diagnosing gastrointestinal tract duplications, but play a role in delineating the nature of the lesion in relation to its adjacent structures. (3,5)

Ultrasonography is generally considered the cornerstone for the diagnosis of an enteric duplication cyst, where this structure appears to have an inner hyperechoic rim corresponding to the mucosa-submucosa layer and an outer hypoechoic rim corresponding to the smooth muscle layer, which is referred to as the well known 'double wall' sign. (1,2,4) This appearance is usually not circumferential as the layers are often nonuniform in thickness, but this doubled-layered wall is found in over 50% of patients. (3) In fact, the diagnostic criteria of enteric duplications include the presence of a common blood supply and signs of double wall and enteric epithelial lining. (5) However, false-positive double wall sign can also be seen with other cystic lesions such as Meckel's diverticulum, ovarian cyst, mesenteric cyst or even sonographic artifacts. (1,4) On the contrary, the Y-configuration sign, caused by the shared muscle layer between the cyst and the bowel, can be more reliable for the diagnosis of an enteric duplication cyst. (1,4,5)
Images for this section:

**Fig. 1:** the 'double wall' sign

© Radiology, P & A. Kyriakou, Children's Hospital - Athens/GR
Fig. 2: tubular duplication cyst

© Radiology, P & A.Kyriakou, Children's Hospital - Athens/GR
Fig. 3: tubular duplication cyst on US and CT

© Radiology, P & A. Kyriakou, Children's Hospital - Athens/GR
Fig. 4

© Radiology, P & A.Kyriakou, Children's Hospital - Athens/GR
Fig. 5: change of shape of the duplication cyst due to peristalsis

© Radiology, P & A.Kyriakou, Children's Hospital - Athens/GR
Findings and procedure details

We retrospectively reviewed the cases of 10 children in a period of 6 years. In all of our cases an abdominal sonogram was performed, for the investigation of various abdominal symptoms, by the same experienced pediatric radiologist with convex 2-5MHz and linear high frequency transducers 10-12 MHz, using gray-scale and Doppler technique. The ultrasound illustrated the presence of a cystlike lesion adjacent to the small bowel loops with a smooth echogenic peripheral wall, consisted of a hyperechoic inner wall and a hypoechoic outer wall. In two of the cases, signs of inflammation were also present such as fluid-fluid level within the cyst and increased vascularity in its wall.

The hypoechoic outer wall of the cystic structure was continuous with the hypoechoic outer wall of the adjacent small bowel, so the Y-configuration was demonstrated on each and every of our cases and the preoperative diagnosis of an enteric duplication cyst was made. All of these children were operated and the presence of an enteric duplication cyst was proven histologically in all of the cases.
Fig. 6: the Y-configuration sign

© Radiology, P & A.Kyriakou, Children’s Hospital - Athens/GR
Fig. 7: the Y-configuration sign in a duplication cyst with internal echoes

© Radiology, P & A.Kyriakou, Children's Hospital - Athens/GR
Fig. 8: the Y-configuration sign in a duplication cyst with internal septa

© Radiology, P & A.Kyriakou, Children's Hospital - Athens/GR
Fig. 9: the Y-configuration sign in a duplication cyst with echogenic material

© Radiology, P & A.Kyriakou, Children's Hospital - Athens/GR
Fig. 10: duplication cyst with fluid-fluid level in keeping with inflammation

© Radiology, P & A.Kyriakou, Children's Hospital - Athens/GR
Fig. 11: increased vascularity in the wall of the duplication cyst in keeping with inflammation

© Radiology, P & A.Kyriakou, Children's Hospital - Athens/GR
**Fig. 12:** the Y configuration sign

© Radiology, P & A.Kyriakou, Children’s Hospital - Athens/GR
Conclusion

Sonography plays a critical role in the workup of pediatric abdominal masses, such as enteric duplication cysts, and often dictates further management. The sonographic double wall sign has been well described in the literature and is often the cornerstone in suggesting the diagnosis of an enteric duplication cyst. However, this sign can also be present in Meckel's diverticulum and pitfalls in relying only on this sign have been described.

The specificity in making the sonographic diagnosis of a duplication cyst increases with the visualization of the split hypoechoic muscularis propria layer between the cyst and the adjacent bowel, resulting in a Y-configuration. This Y-configuration, if visualized in vivo, as it was in our cases, proves to be a key sign in establishing a preoperative diagnosis of an enteric duplication cyst in children. (1,4)
Personal information

Kleanthi Kalogerakou
Radiology resident
Department of radiology
Children's Hospital 'P&A Kyriakou', Athens, Greece
e-mail: cleanthie@hotmail.com
References


